

PRACTICE OF MEDICINE

VOLUME X

PRACTICE OF MEDICINE

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VOLUME X

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INTRODUCTION

In approaching the description of the cranial nerves one is tempted to follow the time-honored precedent and describe them, as to their origin, their course, their distribution and their function, in the usual manner. To do this, however, does not seem quite accurate, for certain changes, readjustments and conceptions are advisable in the consideration of both their anatomy and physiology. In consequence, the author will endeavor in this section to note these anatomical and physiological differences.

The old numerical order used in describing the so-called twelve pairs of cranial nerves will be altogether discarded and an attempt will be made to use the nomenclature which seems the most simple and advantageous. In reviewing the subject, we find that by some odd circumstance the nerves supplying the eye muscles are commonly described as the third, fourth and sixth nerves, with the fifth, a nerve not related to the eye, injected between the fourth and sixth. The old numerical order is supposed to have come into being through the location of the origin of the individual nerve from before backward, but since we have changed our idea of the origin of many of these nerves, and since some have streaks of nuclei of origin rather than points, and these long nuclei pass those of other nerves, it is not well to give them a numerical designation. It seems equally difficult to understand why the old fifth, or trigeminal nerve, is described as one nerve, when in reality there are two distinct nerves embraced in this description—one sensory and one motor—with separate origin, course, distribution and function. In this article, therefore, we shall describe the sensory, trigeminal, and the motor, masseter nerve, separately. Again the old seventh, or facial nerve, is commonly described as embodying the intermediate nerve, or nerve of Wrisberg, as its sensory root, although some have described it as a separate nerve (the thirteenth nerve). Even though it has ganglionic or fiber connection with the facial, the vestibular and the glossopharyngeal, it really appears to be a separate nerve and will here be so

considered. The old eighth nerve, even if the cochlear and vestibular portions are to be considered embryologically the same, will be described, not as the auditory nerve consisting of two parts, but as two separate cranial nerves, the cochlear nerve and the vestibular nerve, for the reason that these nerves are distinctive in their origin, course and function.

There is some fault to be found with the nomenclature of the cranial nerves other than their numerical designation, in that several names are usually given the same nerve. As multiple names are confusing and cumbersome, it seems wise to select what may be considered the best name and let the others become obsolete. Choice of names is always more or less individual, but it would seem that for the cranial nerves there should be either some anatomical or physiological reason for the choice. Thus why should the vagus nerve be also called the pneumogastric, or the trochlear be termed the patheticus, or the trigeminal be termed the trifacial?

The cranial nerves may have motor, sensory, special sense and vegetative functions and, at times, several of these functions belong to the same nerve. When this is so, the fibers for these special functions are frequently carried in one and the same nerve sheath.

Roughly speaking, the cranial nerves supply with their various functions much of the head, including the face, the neck, and to some extent, the viscera.

It seems hardly necessary to mention that in speaking of a cranial nerve in the singular, which is more convenient, the same description applies to its mate, as all of the cranial nerves are in pairs.

In view of what has been said, the cranial nerves will be classified in this article as follows:

Olfactory Nerve: Special Sense—Smell.

Optic Nerve: Special Sense—Sight.

Oculomotor Nerve: Motor.

Trochlear Nerve: Motor.

Abducens Nerve: Motor.

Trigeminal Nerve: Sensory, and Special Sense—Taste.

Masseter Nerve: Motor.

Facial Nerve: Motor.

Intermediate Nerve: Special Sense—Taste (also Secretory).

Cochlear Nerve: Special Sense—Hearing.

Vestibular Nerve: Special Sense—Equilibrium.

Glossopharyngeal Nerve: Sensory, Motor, and Special Sense—Taste.

Vagus Nerve: Sensory, Motor and Autonomic.

Spinal Accessory Nerve: Motor.

Hypoglossal Nerve: Motor.

Thus it may be seen that we have fifteen pairs of cranial nerves, instead of twelve, according to the old arrangement. Even if this departure is radical, the author hopes it will be considered timely and sensible.

Since the first publication of this division of the cranial nerves there has been a tendency toward the adoption of some such classification and in no instance has the author received criticism of their division into fifteen instead of twelve pair.

OLFACTORY NERVE

The olfactory nerve is the nerve pertaining to the sense of smell. Instead of the olfactory nerve being described as having a superficial and deep origin with the fibers running to the peripheral distribution, it will be described, as is sometimes done, as arising in its special sense-organs with its fibers carrying afferent impulses running to the olfactory bulb and thence to the cortical centers. Hence the nerve arises in special receptors for the sense of smell in the Schneiderian membrane of the upper nares, whose filaments, some twenty in number, go through the cribriform plate of the ethmoid, and end in the olfactory bulb, there to arborize with cells of the efferent neurons.

The olfactory bulbs on either side of the brain are situated in the anterior fossæ of the skull, just below the under surfaces of the frontal lobes. From the bulb thus situated at the base of the brain, neurons, forming the olfactory tract, carry the smell impulses to the cortical centers of the sense of smell, which are chiefly situated in the uncinate gyrus, but some of the fibers go on to the optic thalamus.

Functional Tests.—The sense of smell may be roughly tested by the examiner occluding with his finger the nostril not to be tested, and making the patient close his eyes. He then holds before the nostril some non-volatile substance, as ground coffee or tobacco, and notes whether the patient can correctly detect the substance. The other nostril is then tested in the same manner. Volatile substances like perfume, alcohol or ammonia, stimulate the taste-bulbs and may thus be recognized by other means than purely by the sense of smell. The individual capacity for appreciation of odors varies considerably, and may be very accurately estimated by Zwaardemaker's olfactometer, but for practical purposes such refinement is not necessary.

Disorders of the Sense of Smell.—The sense of smell may be hyperacute, diminished, absent (anosmia), or there may be hallucinations of smell.

Increased sense of smell may occur from training, as in tobacco buyers, or it may occur physiologically in certain conditions, as during pregnancy, or even the menstrual period, or it may be increased in hysteria. Smell may also be increased from peripheral irritation of the upper nares. Smell in man has lost much of its function and is more acute in uncivilized than in civilized peoples. In certain lower animals, the hound dog and the deer, for instance, it is very highly developed.

Decreased or absent sense of smell may be congenital, or it may be due to toxemia or mineral poison, or it may occur from local conditions of the upper nose obtunding the nerve terminals, or from lesions of the olfactory tract or bulb. It may also occur from fracture of the nose, or of the anterior fossa of the skull, or from frontal lobe tumor, or, functionally, in hysteria. Atrophy of the nerve, usually of syphilitic origin, may take place and cause an anosmia. It is interesting to note that albinos usually have diminished or absent sense of smell. Their vision is also diminished.

Hallucinations of smell have been noted in tumors affecting the uncinate gyrus. In one of the author's cases, the location of a temporo-sphenoidal tumor was determined chiefly by an hallucination of the smell of blood, of which the patient constantly complained. These hallucinations of smell accompanied by certain other manifestations are frequently called the hippocampal, or uncinate fits of Hughlings Jackson. These consist in a subjective sensation, amounting to an hallucination; usually disagreeable, of smell, or sometimes of taste, or of both, often preceded by an epigastric aura. The muscular movements of smelling, chewing or tasting may occur. They may also be accompanied by salivation, dreamy states and hemianopia.

Some observers think that most *hay-fever*, or rose cold, is of psychogenic origin rather than due to such stimuli as pollen or horse odor. Just how they account for the psychogenic factor acting in a particular month, usually August, the author has not heard explained. It is probable, however, that disgust for certain odors is, at times, due to sexual or psychogenic factors and these factors may have much to do with the disgust for food observed in these patients. It would be interesting to occlude the nostrils in some of these cases and see if it had any influence on the appetite. Hay-fever may be due to a reaction from pollens, or a similar condition occurs from an infection of the upper nasal passages. In the latter cases autogenous vaccine is frequently of great benefit.

It has been suggested by Weir Mitchell that certain phobias, as the *cat phobia*, have origin in the sense of smell. We should remember in this connection that the skunk belongs to the cat family. Not enough work has been done on the physiology and psychology of smell. It would also be interesting to learn what effect the *odor of incense* has upon the devotional attitude, and just what is the relation of smell perception to general anesthesia, or of perfumes to sex attraction stimulation. Certain perfumes are sold for the purpose of sexual stimulation. Certain volatile substances when smelled, ammonia for example, seem to stimulate the circulation. Much of this, of course, is from its stimulating effect on the lungs by irritation, but the author is not at all certain that part of its circulatory stimulation effect is not through some reflex influence from both the taste and the smell-sense.

Mechanism of Smell.—Special smell, or olfactory, cells lie among the epithelial cells of only a small area of the nasal mucous membrane. Probably, as von Brunn believes, they are only to be found in the mucous membrane over the nasal septum and a portion of the upper turbinate bone. The cells are elongated nerve-cells on the free end of which is a tuft of hair-like processes.

Hyglier has shown that smell, like sound and light, travels in waves, and that different odors have different wave lengths. It was formerly thought that odors traveled in particles which impinged upon the special olfactory cells.

That the flavors of many foods, especially certain fruits, and also of wines and cigars, are enjoyed, not through the taste, but through the

olfactory appreciation, is evidenced by the fact that they lose their flavor when the smell-sense is defective from a cold in the head or from occlusion of the nostrils.

OPTIC NERVE

The optic nerve is in reality a brain tract. It is special sense in function and is the nerve of sight. It also contains afferent fibers for pupillary reflex. Its origin is in the cells of the retina which are special sense cells of ganglionic character and their fibers unite to form the optic nerve. These fibers pass backward through the optic foramen of the orbit to form the optic chiasm. Fibers coming from the outer, temporal part of the retina do not cross, but the larger number, which do cross, come from the inner, or nasal, two-thirds of the retina. These two sets of fibers, the crossed and uncrossed, join to form the optic tract which curves around the crus cerebri and form the lateral and mesial roots which connect with the external geniculate body, the anterior tubercles of the corpora quadrigemina and the pulvinar of the optic thalamus (Dana). The fibers then enter the posterior part of the internal capsule and from thence go to their cortical centers in the occipital lobe in the cuneus and about the calcarine fissure. Thus the temporal side of the retina has a neuronie connection with the occipital lobe of the same hemisphere, while the nasal half of the retina has a neuronie connection with the occipital lobe of the opposite hemisphere.

There are many disease conditions connected with the optic nerve and it seems wise to first take up the symptom of hemianopia or half vision, which indicates a lesion, and has definite localizing value in judging the situation of lesions, in the visual pathway at, or behind, the optic chiasm.

Hemianopia.—Hemianopia is a condition of half vision due to blindness of one-half of the retina, this condition not being due to disease of the retina itself. It is measured by charting the visual field of each eye separately.

Lesions involving the optic chiasm are most frequently due to tumors of the pituitary gland, but may be due to tumors in the neighborhood of the pituitary fossa. Syphilitic or inflammatory exudates may also constitute lesions of the chiasm.

If the lesion affects the external tract of the chiasm on either side, it will produce loss of function in the retina of the temporal half of the eye on the corresponding side, or the nasal half of the vision field, and is known as *unilateral nasal hemianopia*. *Bilateral nasal hemianopia* is extremely rare and can only be due to a symmetrical lesion in each cerebral hemisphere affecting both external tracts, which are quite a distance apart, or to two lesions.

A lesion in the center of the optic chiasm will catch the decussating, or crossed, fibers from both eyes and produce loss of function in the cells of the nasal halves of both retinae, or give blindness in the temporal halves of each vision field. This condition is known as *bitem-*

poral hemianopia. A tumor of the pituitary body is the most probable lesion in this location, as the central part of the optic chiasm is situated over the sella turcica. If the central lesion extends to one side,

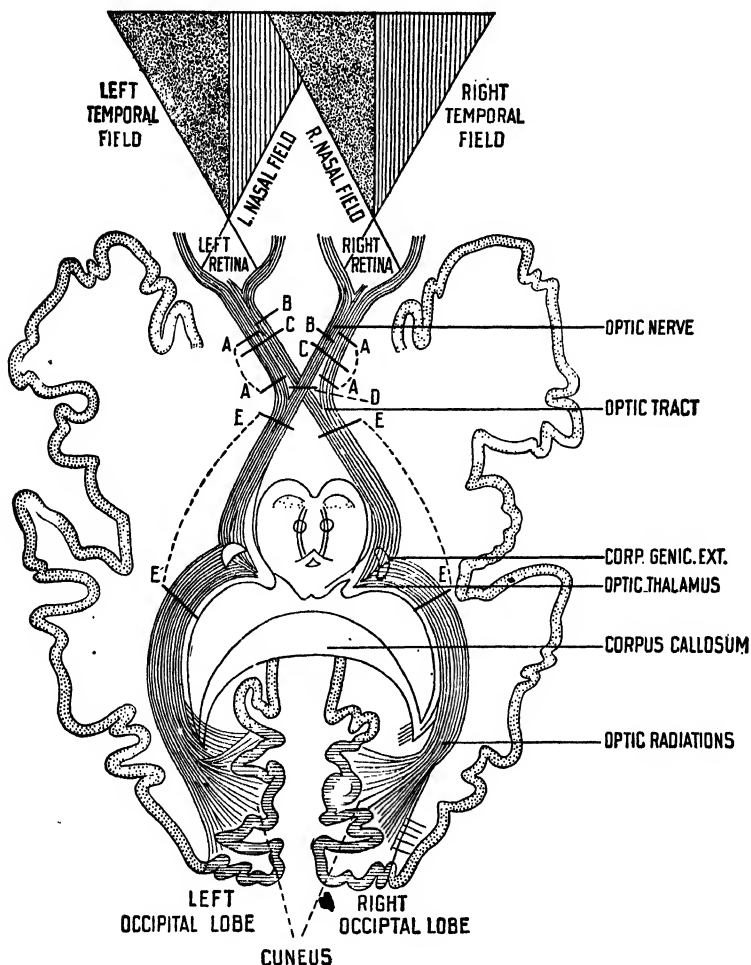


FIG. 1.—OPTIC CHIASM AND TRACTS. (Modified from Violet.)

or the other, it will give total blindness in the eye on the side of the lesion and blindness in the temporal field (*unilateral temporal hemianopia*) in the opposite eye. Of course, if the lesion extends far enough on both sides, it will produce total blindness.

If there is a complete lesion of the optic nerve on either side, anterior to the optic chiasm, it produces total blindness of the corresponding eye. If the lesion affects only the outer half of the fibers of the nerve, *unilateral nasal hemianopia* may be produced, or if the internal half of the fibers only are affected, *unilateral temporal hemianopia* will be manifest.

A complete lesion of the optic tract, posterior to the optic chiasm, will give nasal hemianopia in the visual field of the side of the lesion, and temporal hemianopia on the opposite side. If this lesion is on the left side, the blindness is in the right visual field, and is known as *right homonymous hemianopia*, and if in the right tract, it produces blindness in the left visual field, and is known as *left homonymous hemianopia*. A lesion of the outer part of the tract, posterior to the chiasm, will produce nasal hemianopia in the field of the corresponding eye. If the lesion affects only the inner fibers it produces *hemianopia of the temporal field of the opposite eye*.

It may so happen that only a quadrant of the visual field is blind, and when this occurs it is due to a lesion in the visual centers in the occipital lobe. This is usually spoken of as "*quadrant hemianopia*."

This occurs when there is a lesion of the cortical half-vision center in the occipital lobe. A lesion above the calcarine fissure, which divides the half-vision center in the occipital lobe into an upper and lower part, will cause blindness in a quadrant of the lower visual fields of both eyes. If the lesion is in the left lobe, the blindness will be in the visual field of the right lower quadrants and vice versa. A lesion below the calcarine fissure will cause blindness in a quadrant of the upper visual fields of both eyes. If the lesion is in the left lobe, the blindness will be in the right upper quadrants of both fields, and if in the right lobe, in the left upper quadrants.

WERNICKE, OR HEMIOPIC PUPIL REACTION.—This is a valuable test for brain lesions. It is not present in lesions of the occipital lobes but rather indicates a lesion back of the chiasma, and below, or at the corpora quadrigemina. Patients who show the hemiopic pupillary reaction have hemianopia and the reaction is tested by throwing a fine pencil, or ray, of light on the blind half of the retina when it will be found that pupillary contraction is absent, but the pupil contracts when the light is thrown on the good half of the retina. It is quite a difficult test to make, and few but experienced oculists are capable of making it satisfactorily.

Variations of Visual Acuity.—Visual acuity may be affected by the various diseases of the optic nerve, but more commonly by errors of refraction due to changes in the shape and refractive power of the lens, the shape and transparency of the cornea and the conformation of the eyeball itself. It may also be affected by hemorrhage or exudation into the retina and by conditions affecting the transparency of the vitreous humor.

TESTS FOR VISUAL ACUITY.—Vision is usually tested by means of Snellen's test cards, which are arranged with a series of letters nor-

mally visible at varying distances from 20 to 200 feet. If letters of a certain size are read at 20 feet, the vision is said to be 20/20 or normal, but if letters meant to be read at 100 feet distance are the smallest that can be read at 20 feet, then the vision is said to be 20/100, and so on.

To test the *field of vision*, a perimeter is used for accuracy, and the point at which an object moved toward the direct line of vision, when the eye is fixed at a point straight ahead, comes into view is said to mark the field of vision in that direction. The arc of the perimeter is then changed and the object moved from another direction toward the line of vision when the eye is fixed, as before. This procedure is repeated in all directions till the field of vision is mapped out accurately. The ascertainment of the visual field is valuable both in organic conditions and in hysteria. In either condition the field may be constricted in all directions, but a tubular field, as it is called, is more common in hysteria. Only in organic conditions, and in certain cases of migraine, is the hemianopic field observed. *Central scotoma* or a blind patch in one or both visual fields may be found. In this condition the patient may see an object moved toward his line of vision, then fail to see it and then see it again. By testing in various directions, the blind area may be mapped out as well as the margins of the regular field of vision. An upper temporal slant or contraction in the visual field is indicative of pituitary disorder, particularly of tumor.

The visual field may be *roughly* tested by making the patient face the operator at about arm's length and look with his right eye steadily at the operator's left eye while the operator fixes the vision of his left eye on the patient's right eye. Both the operator and the patient close the eye not to be tested by covering it with the hand. The operator then moves his free hand, or some small object in his hand, toward the line of vision thus maintained, and instructs the patient to notify him as soon as the object is seen. This is repeated from various directions; usually from the temporal side, from the nasal side, from above and below is sufficient. If the operator's field of vision is normal, the patient should see the object as soon as he does and by this means he can judge the extent of the patient's field.

The color fields are of importance and are tested in the same way as the visual fields, only colored objects are moved toward the line of vision.

The physiological order of the color field, from without inward, is blue, yellow, red, green, violet. In hysteria, red is often last to be affected, so that it may be larger than or equal to the blue field. Various other changes in color fields may take place in hysteria and in this condition the perception for green is most easily disturbed.

Impaired Conditions of Disc and Retina Located by Means of Examination of the Fundi.—The appearance of the optic nerve, as it spreads out over the inside of the eyeball at the back, is spoken of as the eye-ground or fundus. It is through the pupil, of course, that the optic nerve is examined, and, except in certain cases of permanent dilatation of the pupil, atropin, homatropin or cocain should be used to dilate the pupil to get a good view of the fundus. In patients over 40 years of age one or two drops of a 4 per cent. solution of cocain is the best of these to use. The examination of the optic nerve, including the disc and the retina, with an ophthalmoscope is one of the most useful means of diag-

nosis in all of medicine and should be mastered by every physician. The disc may be examined to see if *neuritis* is present, which is shown by reddening or swelling of the disc (known as *choking*), or whether *atrophy* is present, as indicated by paling of the disc. Swelling or choking of the discs may represent such serious conditions as *increased intracranial pressure* from whatever cause, *brain tumor*, *brain abscess*, *hemorrhage*, the various forms of *meningitis*, etc. Atrophy of the discs denotes degenerative processes in the central nervous system. The retina may be found to be inflamed from syphilis, nephritis or other causes, and in the retina, especially when it is inflamed, hemorrhages may occur. The age of the hemorrhage may be indicated by its color, the hemorrhagic spot being red when fresh, and darker when older. Hemorrhages of long standing are nearly black. The veins may be engorged and tortuous, or in certain conditions the arteries may be seen to be hardened or almost obliterated. White patches may be seen in albuminuric retinitis. Coal-dust appearing pigment accumulated around part or the whole of the margins of the discs is an indication of syphilis.

Amblyopia.—Amblyopia is a transient blindness due to toxic disturbance, usually from tobacco or alcohol, or to hysteria. It may also occur in connection with diabetes. The amblyopia may be either for vision or color. Color appreciation vision is tested by Holmgren's test wools. This test consists in the matching and comparison of shades of color by means of varicolored skeins of wool. In pronounced cases, the patient usually picks out skeins according to their brightness, regardless of color. Blindness for certain colors may occur in hysteria. The curious fact has been observed that preceding the objective symptoms of multiple, or insular, sclerosis, the history of brief transient blindness may frequently be elicited from the patient.

Optic Neuritis.—Optic neuritis, or papillitis, is an inflammation of the head of the optic nerve as it enters the eyeball. It may be seen in brain tumors, brain abscesses, nephritis, lead poison, tuberculous meningitis, certain infective conditions, and occasionally in polyneuritis. Vision may remain good for a considerable time and there is but little pain or discomfort accompanying the condition. Vision and color field limitation or scotoma may occur. Optic neuritis may be unilateral or bilateral. In the early stages of the condition the disc and retina are reddened. A swelling of the disc takes place, due to serous infiltration, and the physiological cupping of the disc is lost. The margins of the disc in time disappear and it becomes elevated above the surrounding retina. This condition is known as *choked disc* and is a valuable sign of increased intracranial pressure.

Retrobulbar Neuritis.—Retrobulbar neuritis occurs when the lesion affecting the optic nerve is behind the ball of the eye and may be due to syphilis, lead poison, rheumatoid infections, diabetes, or to tobacco or alcohol toxicosis. Changes in the elevation of the disc are absent in this condition, unlike the usual optic neuritis.

Optic Atrophy.—Optic atrophy may follow optic neuritis directly, and, when it does, it is known as secondary optic atrophy and hence may be a sign of gross brain lesion. Primary optic atrophy may occur in multiple or insular sclerosis, in hereditary ataxia, or be inherited, but the usual cause is some chronic form of syphilis of the nervous system.

The pupil becomes dilated and immobile to light reaction, vision is gradually diminished and the visual field becomes concentrically contracted. Sight may be fairly well preserved until late but the condition is never recovered from and is but rarely arrested in its progress. The disc may have a marked, cupped appearance and be of an opaque or grayish color. This paleness of the disc may be uniform or more marked in the outer or inner margins. Pallor of the discs is frequently seen in cases of spastic diplegia from long-continued increased intracranial pressure, as emphasized by William Sharpe.

Retinitis.—Retinitis is an inflammation of the retina which may be caused by many disease conditions, the most common of which are syphilis and nephritis. In these conditions the retinitis is respectively termed syphilitic retinitis or retinochoroiditis, and albuminuric retinitis.

SYPHILITIC RETINITIS.—Syphilitic retinitis may be circumscribed or diffuse. In circumscribed retinitis along the blood-vessels, or near the macular spot, is a light yellowish exudate which is replaced by scar tissue and may lead to detachment of the retina. Scotomata and irregularity may be observed in the visual field, and the irregular whitish remains of exudate or connective tissue may be observed with the ophthalmoscope; also vision may be affected, especially at night (night blindness). The condition may occur in the secondary, but usually in the tertiary, stage of syphilis and is treated by antisyphilitic measures. The retinitis is no contra-indication, but rather an indication, for the intensive use of salvarsan.

ALBUMINURIC RETINITIS.—Albuminuric retinitis is an indication of nephritis in either the acute or chronic stage and is frequently seen in the albuminuria of pregnancy, or in sclerotic kidneys. In albuminuric retinitis irregularly shaped white patches are observed and the disc may be swollen, and hemorrhages into the retina are not infrequent. Fatty degeneration of the diseased area may be observed. The disc may, or may not, be choked or pale. The prognosis is usually not good, except in pregnancy, and the treatment consists in treating the causative condition.

INTRA-OCULAR SIGNS OF DIABETES.—Hyperopia is rare but myopic increase, said to be due to increase in the density of the lens from swelling of the lens, is frequent. The fact that the Hebrew race is subject to both myopia and diabetes is probably coincidental. Eye diseases which are common in diabetic patients are xanthelasma, iritis, cataracts, retinitis and lipemia retinalis.

Visual Aphasia.—Visual aphasia concerns itself with those forms of disturbance with speech and writing due to interference with the visual centers of language and the tracts associated with these centers. The cerebral centers connected with speech occupy a large area of the cerebral cortex and subcortical region, the auditory centers being situated about the junction of the first and second temporal gyri; the motor center occupies the back part of the third frontal convolution; the visual centers of speech are in the angular gyrus; the writing center is in the caudal third of the second frontal convolution; and what is thought to be the naming center, a center for concrete concepts of objects named, is said to be in the third temporal convolution. Besides there are other associated language centers.

IMPORTANT VARIETIES OF VISUAL APHASIA.—*Mind-blindness*, a condition due to cerebral disturbance in which the patient cannot recognize, by sight, once familiar objects. *Mind-blindness* is used to cover what is known as *word-blindness*, or an inability to recognize written or printed words. As the patient cannot recall the visual images he also cannot write, or copy writing or printing. But in certain cases of uncomplicated *mind-blindness*, he can write spontaneously or from dictation, but the moment his eyes are removed he cannot read a word he has written. In *mind-blindness* the patient cannot recognize familiar faces, places, or things in the room and hence is disoriented. In *aphasia* and *word-blindness* the lesion, in right-handed people, is in the left hemisphere. The side on which the lesion is situated in left-handed individuals is frequently doubtful, although in pure left-handedness with transposition of the viscera it may be considered to be in the right hemisphere.

Alexia is an inability to read because of brain disturbance, and in this, the lesion may be variously situated because of the many associations in the zone of language.

Word-dumbness, or so-called optic aphasia, is the inability of a patient with good eyesight to name an object shown him, although he recognizes it and knows its uses. At times although the patient will call this object by entirely the wrong name, he may use a name somewhat associated with it, as, for instance, a patient of the writer called a key, a door. More often, however, he cannot name it at all, but if given, say a key, which he cannot name, he can unlock the door with it. Lesions producing *word-dumbness* may be, like those producing *alexia*, variously situated in the cortical language zone.

Ophthalmic Migraine.—Ophthalmic migraine has many synonyms, common among which are *hemicrania* and *sick headache*. It is most frequently spoken of simply as *migraine*. *Migraine* is an intense, prostrating, periodic headache, usually with an hereditary tendency, which is accompanied by nausea, vomiting and visual phenomena. An attack may last for a few hours to a few days and the attacks may be frequent or occasional in occurrence. There are many abortive forms and the pain may be bilateral.

Etiology.—The cause of the condition is far from settled. Many men of great intellect have been among its victims. One prominent neurologist,* himself a sufferer from the disorder, told the author that he had never, in a long practice, seen a case of migraine in a person who led a real outdoor life. It nearly always occurs in brain workers, or those of a neurotic tendency, and, as a rule, affects several members of a family.

Angioneurotic edema of the brain, arterial spasm of blood-vessels, toxemia, pituitary gland disturbance, and reflex conditions, have all been assigned as the cause of migraine. Spitzer believes it is due to a constitutional anatomical defect causing a partial blocking of the foramen of Monroe. In its suddenness, periodicity and in other respects, migraine much resembles epilepsy.

Symptomatology.—The patient may have a sense of well-being the day previous to the headache, which is frequently followed by yawning, heaviness, vertigo or depression, or he may awaken with the headache,

* The late Dr. John K. Mitchell, of Philadelphia.

or it may strike him suddenly during the day. Sometimes the attack is preceded by unilateral paresthesia or anesthesia. Just what relation migraine has to the optic nerve is unsettled, but the nerve is certainly functionally disturbed. Visual symptoms are frequently early and pronounced and may consist of photophobia, blurring of vision, scotomata, seeing spots, images, zigzag patterns, colors and flashes before the eyes. Sometimes slight ptosis or diplopia will be present and when this is the case, the condition has been spoken of as ophthalmoplegic migraine. The eye symptoms are usually followed by a one-sided intense headache. The headache may, however, be bilateral. During the headache the pupils are usually contracted. The headache is most intense, varying in character in different individuals. The patient is at times delirious. The headache usually leaves suddenly either on the onset of vomiting or not. Nausea and vomiting are common symptoms of the condition and may, or may not, be related to food. Anorexia is often present, but if not, the intaken food is usually ejaculated by vomiting. Throbbing, flushing and various vasomotor disturbances occur.

Diagnosis.—The diagnosis of migraine is made upon the periodicity of the attacks, the visual phenomena, the family tendency, prostration, the gastric disturbance and the intensity of the headache. The chief mistake in diagnosis is made, when the stomach symptoms are severe, by attributing the trouble to gastro-intestinal disease.

Treatment.—The treatment divides itself into the treatment of the attack and the treatment of the individual in general. For the attack, the most satisfactory method in the author's hands has been to put the patient **absolutely to bed**, to **prohibit**, during the stage of nausea and vomiting, **all food by mouth** and to **nourish** the patient, when necessary, **per rectum**. If taken at the beginning, a tablet of **nitroglycerin** dissolved on the tongue and the administration of a dose of **magnesia** frequently aborts an attack. **Pyramidon**, **cafein** and **acetanilid**, and **chloretone** are useful when they can be tolerated. A **mustard plaster** to the back of the neck, and frequently **ice** to the forehead are useful.

Between the attacks the patient may take small doses of **bromids** or **cannabis indica** with benefit. Cannabis Indica is best given beginning with ten drops three times the first day, eleven three times the second, and twelve three times the third, and so on until thirty drops are reached. The **diet** should be **simple** and **nourishing**. **Tobacco** should never be used to excess and **alcohol** should be prohibited. The patient should be in well-ventilated surroundings, **live out of doors** as much as possible and take a full amount of **exercise**. The **eyes** should be properly **refracted** and **overstudy** and **late hours** should be avoided. Pituitary gland extract may be tried. The whole gland in dosage of 2 grains (0.13 gram) three times daily may be used.

Pardee has shown that a bitemporal headache is often due to acute pituitary undersecretion and may be relieved by the administration of pituitary extract. De Schweinitz and Hill have shown that the visual field in these cases shows an upper temporal deficiency.

Prognosis.—The prognosis is disappointing as a rule. The condition frequently disappears in women after the menopause. Because of the indiscretion of physicians, many patients contract the morphin habit, which, of course, only makes matters worse.

OCULOMOTOR NERVE

The oculomotor nerve is motor in function and supplies all the muscles of the eye, except the superior oblique, supplied by the trochlear, and the external rectus, supplied by the abducens. It also supplies the voluntary part of the levator palpebræ superioris (the involuntary part being supplied by the cervical sympathetic) and has fibers which, through the ciliary ganglion and short ciliary nerves, supply the non-striated sphincter pupillæ and ciliary muscles. The nerve has thus to do with the dilatation and contraction of the pupil, and elevation of the eyelids as well as the movement of the eyeball.

The origin of the oculomotor nerve is in the aqueduct of Sylvius and the nucleus of the oculomotor nerve receives fibers from the abducens of the opposite side. The oculomotor nerve comes out in front of the pons and passes the clinoid processes to, and along the outer wall of, the cavernous sinus and divides into two branches which enter the orbit through the sphenoidal fissure below the trochlear nerve. The superior branch passes inward over the optic nerve and supplies the superior rectus and levator palpebræ muscles. The inferior division divides into three branches, one supplying the internal rectus muscle, one the inferior rectus and the third the inferior oblique.

Complete paralysis of the oculomotor nerve will result in ptosis from paralysis of the levator palpebræ, external strabismus, dilatation of the pupil from paralysis of the sphincter fibers of the iris, loss of the power of accommodation, slight exophthalmos due to relaxation of the paralyzed muscles supporting the eyeball, and will cause the eye to look in a downward and outward direction. In the ptosis of oculomotor paralysis, the frontalis muscle overacts, while in hysterical ptosis, or that from myasthenia gravis, it rarely does. Partial paralysis may cause one or more of these symptoms in a greater or less degree. Irritation of the oculomotor nerve causes spasm of the muscles supplied by it. Internal strabismus may occur from spasm of the internal rectus, accommodation for near objects only from spasm of the ciliary muscle, or contraction of the pupil from spasm of the sphincter pupillæ.

The nerve may be affected from lesions pressing on the cavernous sinus, lesions of the bone of the orbit, or sphenoidal fissure, or hemorrhage or new growth in the brain. Syphilitic periarteritis frequently affects it, as the nerve lies between the superior cerebellar and posterior cerebral arteries. The nerve is often affected in locomotor ataxia, by partial or complete paralysis. One of the chief diagnostic symptoms of locomotor ataxia is the Argyll Robertson pupil, which is a pupil that will not contract to light but does to accommodation. This is due, as Marina has shown, to degeneration of the ciliary ganglion. In paralysis of the oculomotor nerve, however, there is immobility of the pupil both to light and on convergence. The pupils are often irregular in syphilis of the nervous system, or they may be dilated in functional, or organic, conditions of the brain, or dilated and fixed from optic atrophy.

Complete paralysis of all the external muscles of the oculomotor nerve may be unilateral or bilateral and is termed *ophthalmoplegia externa*. When bilateral it may occur from lesions of the mesencephalon and cortical oculomotor tracts. Syphilis, dilatation of, or pressure on the third ventricle, tumor and hemorrhage, are the usual causes of oculomotor palsies. A certain form of migraine known as *ophthalmoplegic migraine* may cause paralysis of the oculomotor nerve, and so may exophthalmic goiter, diabetes, and thrombosis, or hemorrhage of the brain.

Hence in paralysis of the oculomotor nerve, the clinical signs are ptosis, elevation of the brow on the side of the paralysis, the eye is turned outward and downward, and the pupil is dilated and fails to react to either light or accommodation.

In exophthalmic goiter there may occur weakness of the internal recti and this is known as *Möbius' sign*. The patient is made to look upward and then at the tip of his own nose. When he attempts this one eye converges while the other eye becomes divergent.

TROCHLEAR NERVE

The trochlear nerve is motor in function and supplies the superior oblique muscle of the eye. This nerve is sometimes called the patheticus, in that the pathetic expression of rolling the eyeballs upward occurs when this nerve is paralyzed. Its origin is in the floor of the aqueduct of Sylvius, just below that of the oculomotor. It runs at first outward, and then inward and backward into the superior medullary velum. Here it decussates with the trochlear nerve of the opposite side, and emerges behind the posterior quadrigeminal body. It passes across the superior peduncle of the cerebellum and around the outer side of the crus cerebri, and passes forward on the outer wall of the cavernous sinus between the oculomotor nerve and the ophthalmic division of the trigeminal, with which it is sometimes blended. It then crosses the oculomotor nerve and enters the orbit through the sphenoidal fissure.

Paralysis of the trochlear nerve makes the patient unable to turn the eye downward and outward, and on an attempt to do this, the other eye is twisted inward, giving the patient double vision or diplopia. One of the early symptoms of affection of this nerve is giddiness on looking downward as on going downstairs. The patient also inclines the head forward and toward the sound side when the nerve is affected.

The trochlear nerve may be, but usually is not, paralyzed alone. Inflammation at the base of the brain, and certain tumors of the base, may affect it in conjunction with the oculomotor and abducens nerves. The trochlear nerve may also be paralyzed from polioencephalitis or from tumors of the cerebellum, the pons or the cerebellopontine angle.

It is thought that ¹ "the connections of the nucleus of the nerve are presumably with the frontal convolutions of the cortex, with the nuclei

of the third and sixth nerves through the dorsal longitudinal bundle, and presumably also with the cochlear portion of the eighth nerve through the superior olive."

ABDUCENS NERVE

The abducens nerve is purely motor in function and supplies the external rectus muscle of the eye. The abducens nerve arises from the upper part of the floor of the fourth ventricle near the midline. A few of its fibers pass to the oculomotor nucleus of the opposite side. The nerve runs on the sphenoid bone, passes through a notch just below the posterior clinoid process, enters the cavernous sinus and then passes through the sphenoidal fissure, to be distributed to the external rectus muscle on its ocular surface. Its course is thus very similar to the oculomotor and trochlear nerves.

Paralysis of the abducens nerve gives internal or convergent squint, in which the eyeball turns inward toward the nasal bridge. In complete paralysis of the abducens there may be partial contraction of the pupil, because some sympathetic fibers to the radiating muscle of the iris pass along the nerve. In cases of long-standing paralysis, the internal rectus from continued overaction becomes contracted and the eye is more or less fixed in the direction of the internal canthus and internal strabismus exists.

In testing paralysis of the abducens it will be found that there is an inability to turn the eye outward beyond the midline and diplopia occurs on an attempt to look outward. The muscles may be attacked by rheumatic myositis (Purves Stewart) but the nerve is more often paralyzed from syphilis of the nervous system than from anything else.

A nuclear lesion of the abducens in the pons will give weakness of the external rectus of the same side and of the internal rectus of the opposite eye through the fibers passing to the opposite motor oculi nucleus. A lesion in the pons may also give facial paralysis of the same side, for the facial root hooks around the abducens nucleus in the pons.

In cases of syphilitic paralysis of the abducens nerve, the condition frequently recovers with the administration of strychnin in addition to the usual antisyphilitic remedies. Strychnin, when given for this condition, must be pushed. The dose may be gradually run up from a thirtieth of a grain, three times a day, to one-tenth, or even one-fifth, of a grain three times a day. The paralysis is practically always unilateral when of syphilitic origin. Abducens paralysis on one side is not an uncommon accompaniment in locomotor ataxia, and when present adds considerably to the patient's difficulty of equilibrium. When both nerves are weakened, or paralyzed, a condition commonly called "cross-eyes" or squint exists. If this is congenital, or occurs in very early life, it is frequently due to gross error in refraction and

not infrequently disappears when the proper glasses are worn. The condition may come on from violent spasms of coughing in cases of whooping cough, or from convulsions.

CONSIDERATION OF OCULOMOTOR, TROCHLEAR AND ABDUCENS NERVES

Certain affections of the oculomotor, trochlear and abducens nerves will now be considered, as these nerves supply all of the external and some of the internal muscles of the eye.

Internal Ophthalmoplegia.—If the ciliary ganglia, or their fibers, or the short ciliary nerves to the iris and ciliary muscle, are alone affected, the condition is termed internal ophthalmoplegia. In this, the pupil is dilated and will not react to light or to accommodation. It may be unilateral or bilateral and is classed with the third nerve palsies. The condition may occur transiently after diphtheria.

External Ophthalmoplegia.—As the oculomotor and the trochlear nerves arise in the floor of the sylvian aqueduct and the abducens has fibers connecting with the oculomotor nucleus in this area, a lesion here may affect both of them partially, or completely, and the condition is known as nuclear, or external, ophthalmoplegia. Usually both eyes are affected. At times, the paralysis is not complete and lateral movements are possible. It may be, but usually is not, associated with internal ophthalmoplegia.

Total Ophthalmoplegia.—When external ophthalmoplegia is associated with the internal variety, it is known as total ophthalmoplegia. When the ophthalmoplegia is total, the patient's pupils are fixed and the eyes cannot be moved in any direction, and reflex nystagmus—both optic and vestibular—is lost. The eye muscles may also be paralyzed in progressive poliomyelitis superior.

Muscular Involvement.—In a destructive lesion at, or above, the internal capsule, the patient cannot turn both eyes toward the contralateral side, so the antagonistic muscles draw the eyes toward the side of the lesion, giving what is known as *conjugate deviation*. When one eye is directed upward and outward and the other downward and inward, the condition is known as *skew deviation* and is due to a lesion of the lateral lobe or middle peduncle of the cerebellum.

When the eye muscles do not work in harmony *diplopia*, or double vision, results. Involuntary rhythmic tremors of the eyeballs, known as *nystagmus*, may occur from weakness of the muscles, or as a symptom of certain brain lesions, especially multiple sclerosis and cerebellar disease. Nystagmus may also be congenital.

The eye muscles can be tested by making the patient, with the head in a fixed position, follow with his eyes the examiner's finger moved in various directions. The excursion of the eyeball is then watched, and any lagging movement and the direction of the lagging is noted. By the same test nystagmus may be observed if present. For fine muscular

imbalance and diplopia, the patient is tested with lights and prisms, according to the methods employed by oculists.

Some of the conditions mentioned need further elaboration, and we shall consider a few of them.

INTRA-OCULAR MUSCLES AND THE PUPIL.—The iris is muscular in structure and has the property of dilatation and contraction which vary the size of the pupil so that more, or less, light may enter the eye. This dilatation or contraction may be accomplished by variation in the amount of light thrown into the pupil, or by the act of accommodation or convergence of the vision on a near object. The iris consists of both circular and radiating fibers. The former are innervated by the oculomotor nerve and the latter by the sympathetic. Between the choroid and the iris is an unstriped band of fibers known as the ciliary muscle which aids in accommodation, and consists of longitudinal and circular fibers, innervated by the oculomotor nerve and the short ciliary nerves from the ciliary ganglion.

The pupils in health are usually equal in size, round in outline, and react to both light and accommodation, but we should remember that slight inequality, slight irregularity and even slight sluggishness to light are not at variance with normal health. However, pupils decidedly irregular in shape and outline, and unequal in size, usually indicate syphilitic involvement of the central nervous system.

Dilated Pupils.—The pupils are usually larger in blonds than in brunets. They are frequently dilated in the nervous, the tuberculous, the anemic, and are commonly dilated in cases of optic atrophy. Dilatation of the pupil is known as *mydriasis* and may occur with the use of certain drugs, as atropin or cocain. Paradoxical dilatation, that is, a condition in which the pupil dilates instead of contracts on convergence, may occur. Under strong emotion, *psychic dilatation* of the pupil may take place. This is not infrequent in fear states, sexual exhilaration and periods of intense interest. Mydriasis is due either to stimulation of the dilator or paralysis of the sphincter fibers.

Contracted Pupils.—Contracted pupils, or myosis, may occur with certain drugs as in morphin habit, and in some pathological conditions such as pontine hemorrhage, and sometimes in syringomyelia, and is either due to stimulation of the sphincter pupillæ or paralysis of the dilated fibers.

In both dilated and contracted pupils the light and accommodation reflex may, or may not, be present.

Argyll Robertson Pupil.—The Argyll Robertson pupil is one that will react to accommodation but fails to react to light. It may be seen, in rare instances, in chronic alcoholism and syringomyelia, but it is nearly always indicative of locomotor ataxia or general paralysis of the insane. The reaction to light is tested by the operator covering both eyes of the patient and then quickly uncovering one of them so that either strong daylight or artificial light will fall upon the pupil and then observe the contraction, if present. The artificial light is best, because accommodation may come into action, thrown on the side of the pupil rather than

straight into it and the test is best made in a darkened room. Reaction to accommodation is tested by making the patient look at a distant object and then quickly focus on a near object, or it may be done by bringing an object from a distance of several feet up toward the patient's eyes and observing his pupils contract as they converge.

Wernicke, or Hemipic, Pupil.—The Wernicke, or hemipic, pupil is one which fails to react to a ray of light thrown on the blind side of a hemianopic eye, but which reacts when the ray is thrown on the non-affected side of the retina. It is a sign difficult to elicit, but it is a valuable one in the diagnosis of certain brain tumors.

Hippus.—In certain cases of hysteria a condition known as hippus may occur, in which the pupils alternately dilate and contract under uniform illumination. All pupils have a state of "pupillary unrest" if observed under high magnification and when this is absent it is a sign of organic disease, but in hippus the excursion is widely greater and can be readily observed with the naked eye, which cannot be done in pupillary unrest.

Accommodation and Light Reflex.—Reaction to light and accommodation may be preserved in oculomotor paralysis and in some cases of atrophy. Loss of accommodation reflex and preservation of the light reflex may be present in certain cases of postdiphtheritic paralysis.

NYSTAGMUS.—Nystagmus is a symmetrical, involuntary oscillation of the eyeballs and is usually bilateral. To test for nystagmus, the patient, with both eyes open, is made to follow with his eyes the operator's finger to one side and then to the other, then upward and then downward and sometimes in a circular manner. When the eyes are undergoing these excursions, especially horizontally at the limits of excursion, nystagmus will manifest itself by the oscillation or jerking of the eyeballs in a rhythmic or pendular movement. If rotary nystagmus is present, it may occur with the eyes looking forward, or when the operator's finger is moved in a circle.

If the ear is syringed with hot or cold water, a nystagmus is set up which is termed vestibular nystagmus. It is said that cold water will produce vestibular nystagmus in the opposite direction from that produced by hot water. (Scott,² quoted more fully by Purves Stewart.³) Nystagmus may also be produced by certain manipulations of the ear and certain ear diseases. It may also be produced by rapidly whirling the patient around, as on a revolving stool.

Nystagmus may be present in certain wasting diseases and in fatigue. At times it is congenital. It is not infrequently seen in albinos. Nystagmus occurs at times in miners and in them is usually vertical.

Nystagmus is a valuable sign in the diagnosis of certain organic nervous diseases, especially disseminated sclerosis, Friedreich's ataxia, cerebellar disease, and sometimes in syringomyelia, hydrocephalus and alcoholic neuritis. It should not be forgotten that nystagmus may be congenital.

DIPLOPIA.—Diplopia is a condition of double vision caused by imbalance, or paralysis of the ocular muscles. It may be produced volun-

tarily, but when involuntary is usually a sign of organic disease. The more acute the diplopia, the more annoying it is to the patient, for in long-standing and congenital cases the patient may involuntarily throw one eye out of use and accustom himself to use the retina of the other eye. When the diplopia is known to the patient he sees double, that is, sees the object, and its image separated from it, on attempting to focus on an object, and the diagnosis is easy for he tells you himself, or when asked, of his double vision. In other cases we have to test for diplopia. The simplest way of doing this, and for diagnostic purposes quite a satisfactory way, is to direct the patient to look at the finger of the examiner with both eyes as the finger is moved, and at certain places the finger will be seen double. If a more elaborate test is desired the examination is made by an oculist with lights, colored glass and prisms. In this way the exact degree of diplopia may be estimated. Purves Stewart³ gives a rule worth remembering—"In all cases of ocular paralysis the affected eye is displaced in a direction opposite to the direction of the traction of the paralyzed muscle, whilst the false image, seen by the affected eye, is displaced in the direction of traction of the paralyzed muscle."

PARALYSIS OF INDIVIDUAL EYE MUSCLES AND THEIR RELATION TO DIPLOPIA.—*The external rectus* may be paralyzed by a lesion of the abducens nerve at any point in its course and produces convergent squint and diplopia. When the external rectus is paralyzed the internal rectus overacts and may become contracted, thus increasing the squint. The eyeball, in external rectus paralysis, swings to the inner canthus of the eye. *The internal rectus* is affected in oculomotor palsy and produces divergent squint and diplopia. The eye tends to swing to the outer corner of the palpebral fissure. Isolated paralysis of this muscle is rare. *The superior rectus* is also affected in oculomotor paralysis and is so rarely paralyzed alone, that its separate paralysis need hardly be considered. Diplopia may be present when this muscle is paralyzed. *The inferior rectus* may be paralyzed and the ball is turned and rotated outward, due chiefly to the action of the superior oblique. It is also rarely paralyzed alone and is supplied by the oculomotor nerve. Diplopia is present in an attempt to look downward. *The inferior oblique* is also supplied by the oculomotor nerve and when paralyzed, the patient cannot look upward and outward. Diplopia is usually present. *The superior oblique* muscle of the eye is supplied by the trochlear nerve and when paralyzed the eye rotates inward and there is diplopia on looking downward.

Exophthalmos.—Exophthalmos consists of protrusion of the eyeball forward through the palpebral fissure. Exophthalmos may be due to a number of causes, probably the most common of which is a thyroid toxicosis known as exophthalmic goiter. Exophthalmos is not infrequently seen in brain tumors, sometimes in certain forms of cerebral myelitis, and at times is due to general debility when the ocular muscles are weak. When the latter is the case, the exophthalmos is said to be

increased when the patient stoops over. Exophthalmos is sometimes congenital, and when so, usually has a familial tendency. Congenital exophthalmos is especially common in the negro race.

The exact mechanism of exophthalmos is not known, but the ocular protrusion is usually supposed to be due to venous congestion of the orbit.

Exophthalmos occurs in varying degrees. It may be so slight as to escape casual detection, or it may be so great that the eyeball may become dislocated through the palpebral fissure and have to be replaced by manipulation. The writer reported two such cases in the *Journal of Nervous and Mental Disease*, June, 1907. One of these instances occurred in a case of cerebral gumma and the other in a case of exophthalmic goiter. Exophthalmos is usually bilateral but may be unilateral.

Various eye signs have been used in the detection of exophthalmic goiter. *Von Graefe's sign* is an inability of the upper lids to follow the movements of the ball downward. It is usually but not always a sign of exophthalmic goiter. *Dalrymple's sign* is a staring expression of the patient due to a widening of the palpebral fissure. *Stellwag's sign* refers to infrequency of winking. *Möbius' sign* is an inability of the eyeballs to properly converge.

The eye muscles in exophthalmos may suffer from slight weakness, with limitations of the eye excursions in various directions, to actual palsy of the extraocular muscles. This eye muscle palsy may occur independent of the usual cranial nerve paralysis. Diplopia is rare in exophthalmos.

TRIGEMINAL NERVE

The trigeminal nerve has formerly been described as the fifth, tri-facial or trigeminal nerve, consisting of a sensory and a motor portion. In this section we will consider the trigeminal as a separate sensory nerve, and the masseter, or old motor branch, as a distinct motor cranial nerve. This motor nerve is not a branch of the sensory nerve, and the origin, course, function and distribution of the two nerves are quite distinct.

The trigeminal nerve is a sensory nerve supplying sensation to the skin and mucous membrane of the face. Its sensory roots terminate in a long tract of nuclei in the medulla, which is continuous below with the substantia gelatinosa, and is called the lower sensor nucleus, and also in cells in the upper sensor nucleus in the floor of the fourth ventricle to the outer side of the nucleus of the masseter nerve (the old motor portion).

On the petrous portion of the temporal bone near its apex, is situated the gasserian ganglion from which two large roots, known as the posterior roots, go to the nuclei in the medulla after entering under the side of the pons near its upper border. The gasserian ganglion gives a few filaments to the tentorium and the dura, and divides into three

large branches, the ophthalmic, the superior maxillary and the inferior maxillary.

The ophthalmic branch runs forward along the outer walls of the cavernous sinus, enters the orbit through the sphenoidal fissure, and breaks into branches to supply with sensation the cornea, conjunctiva, iris, lachrymal gland, part of the nasal mucous membrane, the skin of the upper part of the nose and the forehead as high as the vertex.

The superior maxillary, or second division of the trigeminal nerve, leaves the skull through the foramen rotundum, crosses the sphenomaxillary fossa, enters the orbit through the sphenomaxillary fissure, and

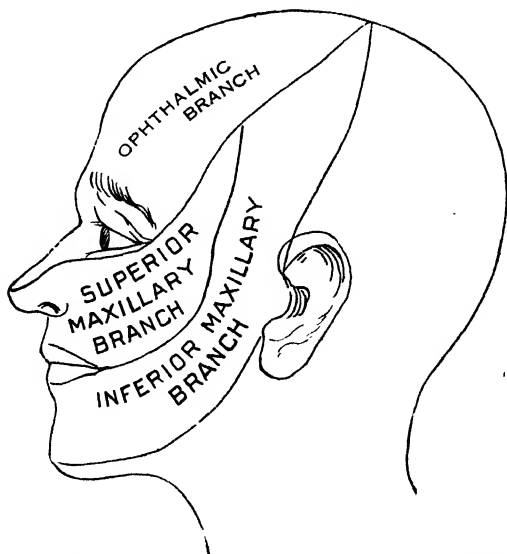


FIG. 2.—APPROXIMATE CUTANEOUS DISTRIBUTION OF THE TRIGEMINAL NERVE.

appears on the face through the infra-orbital foramen in the malar bone. It gives off recurrent branches to the dura and divides into branches which supply sensation to the skin of the side of the nose, the upper lip, the upper part of the cheek, part of the temple and the lower lid. It also supplies the upper teeth, the mucous membrane of the upper gums, the upper buccal region, the upper lip, the uvula, tonsil, nasopharynx, the lower part of the nose, the middle ear and some taste fibers.

The inferior maxillary, or third branch of the trigeminal, leaves the cranium through the foramen ovale with the masseter nerve. It also gives filaments to the dura and divides into branches which supply the lower teeth, the mucous membranes of the tongue as far back as the circumvallate papillæ (Purves Stewart), the gums, the floor of the mouth, the lower part of the cheek and the salivary glands. It sup-

plies the skin of the posterior part of the temple, part of the pinna, the anterior and upper wall of the external auditory meatus and the anterior part of the drum, part of the cheek, the lower lip and the chin. Its lingual branch, in conjunction with the corda tympani, has to do with supplying taste to the anterior two-thirds of the tongue.

Paralysis of the trigeminal nerve gives anesthesia of that half of the face supplied by the nerve from near the lower border of the jaw to the vertex, and includes the mucous membrane of the parts supplied. Paralysis of any of the branches of the nerve gives anesthesia to the parts supplied and may also, if certain branches are paralyzed, affect taste, lacerimation and salivation. As the deep sensation of the facial muscle is also supplied by the trigeminal, there is awkwardness and slight pseudo-palsy of that side of the face.

Irritation of the trigeminal nerve produces pain. This may be central or peripheral in origin. The nerve is not infrequently affected by brain tumors, especially in the temporosphenoidal region. *Tic douloureux*, or trigeminal neuralgia, or neuritis, is a severe affection of one or more branches of the nerve and will be described later, as will also a certain form of head pain due to neuritis of the dural branches of the trigeminal nerve.

The trigeminal nerve has to do with smell, for partial anosmia takes place when the upper branch is rendered anesthetic; it has to do with taste through its corda-tympani connection; and with hearing through its distribution to the ear. The salivary and lacrimal secretions may be affected by injury to the nerve, which, with certain trophic disturbance, may cause loss of the eyeball or infection of the mouth and injury to, or loss of, teeth. Considering all of these disturbances and remembering that the trigeminal is the great nerve of touch sensation to the face, including the mucous membranes of the eyes, nose and mouth, we can imagine its importance.

Progressive Facial Hemiatrophy.—This is a disease which usually appears before puberty and is said to be more common in females than in males. The skin becomes atrophic and then wrinkled over the affected side of the face. The condition spreads and the muscles and bones, and even the tongue on that side, become atrophic. There is no reaction of degeneration, paralysis or sensory change, and the tongue protrudes straight. The beard may be affected and turn white or fall out. Practically nothing is known of the cause of the disease except that it is supposed by some to be due to an interstitial neuritis of the gasserian ganglion, and it is sometimes a symptom of syringobulbia. Treatment is without avail.

Injury of Trigeminal Nerve in Fracture of Base of the Skull.—In fracture of the base of the skull the trigeminal nerve has been injured, and when this occurs anesthesia of the part supplied takes place.

Trigeminal Dural Neuralgia.—*Etiology.*—In the last ten years the author has observed a type, or variety, of head pain, neuralgic in character, yielding but slightly to the usual analgesics, having more or less distinct manifestations and due, in his opinion, to neuralgia, or

neuritis, in the *dural* branches of the trigeminal nerve, just as *tic douloureux*, with which it is at times associated, is due to neuralgia in its peripheral branches. For this condition the name of trigeminal dural neuralgia is selected.

The dural branches of the trigeminal nerve spring from the gasserian ganglion, the sensory roots, or the nerve trunks themselves. Dana, in his text-book, gives the dural distribution of the trigeminal nerve as follows: "It gives sensation to the anterior three-fourths of the dura mater, the falx and probably the tentorium. The pia and arachnoid are not sensitive. The posterior fossa and the occipital part of the dura mater are supplied with the vagus. The trigeminus also supplies the above named parts with trophic, vasomotor and secretory fibers." As the brain itself is insensitive, then intracranial head pain must be due to involvement of the sensory nerves of the dura.

The cause of trigeminal dural neuralgia seems to be the same as in trigeminal neuralgia, which is usually constitutional. These causes are most frequently malaria, focal infections, diabetes, anemia, syphilis, arteriosclerosis, or sometimes other general disturbance.

It is needless to say that the cause should be most diligently searched for and, as far as possible, corrected, for, in the treatment of the causative factor lies the hope of the relief of the neuralgia.

Symptomatology.—The nature of trigeminal dural neuralgia is that it occurs in attacks of sharp, intense pain in the head of irregular periodicity; that nausea, vomiting, flashes of light and familial tendencies are not prominent features as they are in migraine; that the pain, as a rule, is unilateral and may radiate to the face, neck or shoulders; that the pain does not yield to the usual headache remedies; that vasomotor and trophic disturbances are common; that this head pain cannot be traced to the ordinary causes of headache producing changes in cerebral circulation, but that it appears to be due to chronic systemic disease; and that remedial medication directed toward these conditions usually cures the dural neuralgia. The author⁴ reported nine cases in 1913 and since that time has seen many others.

Pathology.—The pathology, like that of trigeminal neuralgia, is probably scant. We only know that in this, as in trigeminal facial cases, there may be found some cell changes in the gasserian ganglion and sometimes evidence of arteriosclerosis in the arteries supplying the nerves.

Tic Douloureux or Trigeminal Neuralgia.—The names *tic douloureux*, trigeminal or trifacial neuralgia, are applied to pain in the distribution of the trigeminal nerve, which comes in spasms, causing a tic, and is due to irritation, or inflammation of the gasserian ganglion, or possibly, in mild cases, of the nerve trunks.

The condition rarely occurs before the age of 35 years, and is most frequently seen in those of advanced years. Fuchs, of Vienna, thinks that this is perhaps due to an impoverished condition of the nerve and its centers, influenced by existing arteriosclerosis.

Etiology.—The cause of *tic douloureux* is usually systemic, that is,

secondary to some constitutional disease. Malaria is a frequent cause and the name brow-ague is popular in malarial districts for neuralgia of the ophthalmic branch. Infective conditions from the tonsils, teeth, sinuses, or elsewhere may cause the condition; anemia, diabetes, arteriosclerosis, and syphilis may have a definite causative relation.

Symptomatology.—The pains of trigeminal neuralgia are described as shooting, boring, burning, aching, tearing or sharp, and usually begin periodically with complete intermissions. The attacks come closer and closer together with greater severity, till the pain is constant, with frequent exacerbations. Once the condition is manifested, a neuralgic habit tends to become established. Many patients develop the morphin habit in their search of relief.

Weather rarely influences the pain, but the patient may wrap up the head to keep a draft from striking the face, as it may increase the pain, so also may chewing, or movements of the face muscles, as in laughing. The pain may involve any, or all, of the branches of the nerve and herpes zoster may occasionally appear. The disease may last for years and the severity be so great that the patient would sincerely rather be dead than alive.

Diagnosis.—One might think the diagnosis easy, but it is, however, fraught with danger. Functional facial spasms are sometimes accompanied by pain. Weisenberg⁵ has suggested that alcoholic injection will aid in the diagnosis, as functional pain persists after the distribution of the nerve injected is made anesthetic. Weisenberg has also called attention to the important fact that neuralgia of the glossopharyngeal may simulate that of the trigeminal because the lower part of the trigeminal and the upper part of the glossopharyngeal intermingle in their sensory distribution. Glossopharyngeal pain is deep in the lower jaw and there is discomfort in the back part of the tongue and throat. Pain from infection of the frontal sinus or antrum may be mistaken for tic douloureux. In all cases these conditions should be eliminated by transillumination or radiography and a careful examination of the nasal cavity.

Tooth root abscesses may cause pain in part of the trigeminal distribution; but this is usually not accompanied by a tic nor is the pain of such long duration as in true tic douloureux. Here again the radiograph comes to our aid. Many good teeth have been extracted in the vain hope of curing tic douloureux. It is useless to extract teeth which the radiograph shows in good condition.

We must also exclude, in the diagnosis of tic douloureux, such conditions as fracture of the base of the skull, brain tumors, or abscesses, or cranial bone infections, which may cause pain in the trigeminal distribution.

Treatment.—The treatment of tic douloureux should be undertaken in stages. The author wishes to again emphasize, as he did in 1908 and 1909,⁶ the value of **medicinal treatment** in these cases and to advocate it before alcoholic injection or surgical intervention is attempted. Natu-

rally the treatment of tic douloureux divides itself into three stages—medicinal treatment, treatment by injections and surgical treatment.

(a) *Medicinal Treatment*.—The chief remedies that are usually relied upon to relieve violent pain are morphin, coal tar preparations or cocaine. These, in any given case, have in all probability been tried. The author makes bold to say that not one of these, except the **salicylates**, is indicated in the treatment of tic douloureux, because they have not proved effective and because they are injurious in their results if continued for any great length of time.

It is quite insufficient, as far as the patient is concerned, simply to make the diagnosis of tic douloureux, but the true cause must be diagnosed in each individual case and treatment applied directly to the condition which underlies the pain. This requires time, patience, laboratory and other tests, a complete history and a careful study of each case.

The **blood should be examined**, and if plasmodium or changes indicative of chronic malaria are found, these, with the history of past malaria, or residence in a malarious district, lead us to a diagnosis of malaria as the cause, and the chief drugs used to combat our tic douloureux become **quinin and arsenic**.

If anemia is found **iron and arsenic** must be pushed.

If in conjunction with our physical examination we find evidence of past or present foci of infection and the history of arthritis, we use the **salicylates** in large doses and remove the infective foci if possible.

We **examine the urine** carefully, and if diabetes or nephritis is present, these conditions must receive our energetic attention. The author has been able to readily control, and keep controlled, the majority of cases of tic douloureux by medicinal treatment.

A word as to the administration of some of these drugs to get the best effect. **Quinin** is best given in solution and not in pills, and most patients can take from 20 to 30 grains (1.3 to 1.95 grams) daily, without disagreeable effects, for a considerable time. Of the salicylates, the author prefers the **sodium salt**, giving it in 10 grain (0.65 gram) doses combined with **sodium bicarbonate** every hour or two for a few days, and then reduces the dose to 10 grains three times daily. **Castor oil** may be given in doses from $\frac{1}{2}$ to 1 ounce (15 to 30 c.c.) daily for several weeks. Purging ceases after the first few doses and the castor oil seems in itself to have a pain-relieving effect.

As adjuvants to treatment, some of the following are useful in lessening the pain: **strychnin**, as recommended by Dana, beginning with $\frac{1}{30}$ grain (0.00216 gram) under the skin and increasing gradually to $\frac{1}{5}$ grain (0.01296 gram) daily; **cannabis indica**, 10 drops of the tincture three times daily and increasing to 30 drops; or **fluidextract of gelsemium** on the ascending scale from 3 to 12 drops. Painting the skin and mucous membrane with a **weak solution of iodine**, or the application of a **hot salt bag**, are good external aids.

(b) *Treatment by Injections*.—Various substances have been used for injection of the trigeminal nerve, but the most satisfactory seems

to be an eighty-fifth to ninetyeth per cent. **alcohol solution** with 4 grains (0.26 gram) of **cocain** to the ounce.

Superficial injection may be made by injecting at the supra-orbital, infra-orbital or mental foramen, but the relief of pain is usually transitory and not very satisfactory.

In using the deep injections it is unsatisfactory and dangerous to inject the superior, or ophthalmic, branch. The other two branches may be injected by several methods. The method of Hugh T. Patrick is simple and satisfactory, and consists in using a straight, sharp needle marked off in centimeters up to 5, and measuring 10 in length. It has a stylet whose blunt edge is even with the end. After the foramen is reached the stylet is withdrawn and alcohol injected. Patrick says, "In injecting the middle branch the line of the posterior border of the ascending orbital process of the malar bone is prolonged to the lower border of the zygoma, and the needle inserted .5 centimeter posterior to this point. It is directed vertically to the anteroposterior line, but inclined slightly upward in a direction which would attain, at the depth of the foramen rotundum, the level of the inferior extremity of the nasal bones. At a depth of 5 centimeters the nerve is reached at its emergence from the foramen rotundum into the pterygomaxillary fossa.

"For the inferior branch the needle is inserted at the lower border of the zygoma, 2.5 centimeters in front of the descending root of the zygoma, which always can be felt, and almost coincides with the anterior border of the external auditory meatus. The needle is directed slightly upward so as to hug the base of the skull, and a little backward, and at a depth of 4 centimeters should reach the nerve at its exit from the cranium."

(c) *Surgical Treatment*.—Operative procedures may be divided into two classes: those in which the nerve is cut after its exit from the skull, and those in which the skull is opened and part of the Gasserian ganglion removed or the sensory roots cut. It is unnecessary to discuss the *modus operandi* of these operations. In neither class have operations on the whole been eminently satisfactory. In the former, results have not always been permanent, and sometimes as many as four operations have been performed upon the same patient, and in the latter, extensive anesthesia is substituted for pain and distressing trophic disturbances may occur.

The Gasserian ganglion operation was introduced by Rose, in 1888, and later modified by Sir Victor Horsley and others, especially Cushing. The operation proposed by Spiller and Frazier of division of the sensory roots above the ganglion may be less dangerous to life.

Turk collected 201 operations on the Gasserian ganglion showing a mortality of 17 per cent. Of the recoveries 93 per cent. were considered cured, leaving 7 per cent., in which the result was not satisfactory. This makes about 24 per cent. of these operations direct failures. The danger of the operation has been lessened in recent years and the results are more satisfactory. Reviewing the function and distribution of the

fifth nerve, we may readily see why some of the following results of the operation may and do occur: Insensitive areas of the skin and mucous membrane render trauma and infection liable; trophic keratitis and other eye injuries occur and even loss of an eye has been known; lessened saliva and partial loss of the sense of taste cause poorer mastication and digestion of food. Anesthesia and dryness of the nasal mucous membrane is another disadvantage. From trophic disturbance, the gums may ulcerate and the teeth may loosen and come out. Thus we see that both the alimentary and respiratory tracts are laid more open to irritation and infection.

When operation becomes the means of relief the radical operation is upon the whole preferable to peripheral attempts.

Frazier,⁷ who had operated on 121 ganglions, advises the avulsion of the sensory root as suggested by Spiller. He operates with the patient in the sitting posture. He approaches the ganglion from an incision allowing him to do so from the middle of the zygoma and severs the posterior sensory roots. With this operation he has had 4 deaths. He considers this operation superior to peripheral operations or alcoholic injections.

MASSETER NERVE

The masseter nerve, formerly described as the motor root of the trigeminal nerve, is purely motor in function. It arises chiefly from cells in the floor of the fourth ventricle, which receives fibers from the cortical cells of the lower part of the central convolutions and runs forward, passing under the Gasserian ganglion, having no connection with it, and leaves the skull through the foramen ovale in the sphenoid bone, through which also passes the inferior maxillary branch of the trigeminal nerve. It is distributed to the masseter, temporal, both pterygoid, the tensor tympani, mylohyoid and anterior belly of the digastric muscles. There is no real connection between the sensory trigeminal nerve and the motor masseter nerve, although the masseter joins the inferior maxillary branch of the trigeminal for a short distance soon after they both pass through the foramen ovale.

Whether the masseter nerve is functioning or not is easily tested by the operator making the patient perform the movement of biting, and observing, or feeling, the masseter or the temporal muscle contract. Paralysis of the masseter nerve gives atrophy of the muscles supplied by it and also reaction of degeneration. Above and below the zygoma there is a hollowing of the face. Upon opening the mouth the lower jaw deviates to the affected side. This, as Purves Stewart notes, is due to paralysis of the external pterygoid, which fails to draw the condyle forward on the affected side.

In the condition known as *pseudo-bulbar palsy* the lesion is cortical and bilateral. In this the jaw hangs open and the teeth cannot be closed; the jaw cannot be moved forward and the lateral movements are inter-

ferred with. In this condition food falls from the mouth unless it can be shoved back with the fingers and often food gets into the nasopharynx. The condition is not infrequently encountered in cases of chronic encephalitis. The jaw-jerk is increased but there is no atrophy or reaction of degeneration.

In tetany, tetanus and strychnin poisoning, there may be a spasm of the muscles of mastication. Convulsive movements of the jaw may occur in epilepsy.

Chattering and grinding movements of the jaw may occur, and, when present, are due to cortical irritation of the masseter nerve. These movements of the jaw are frequently seen in paralysis agitans and the mechanism of them is not quite clear. Irregular movements of the jaw may occur from hysteria, dementia præcox and multiple sclerosis. Clamping of the jaw may be due to hysteria, inflammation, tetanus (lock-jaw), or may occur in certain mental conditions as dementia præcox and manic-depressive insanity. White and Jelliffe⁸ believe that irregular movements and clenching of the jaw are frequently psychogenic in origin. Any of these toxic or clonic spasms of the jaw are known as trismus. Peripheral paralysis of the masseter is rare and when it does occur it is usually due to trauma.

The masseter may be paralyzed from a lesion in the nucleus. The paralysis may be unilateral or bilateral. The muscles supplied by the nerve are flabby in nuclear lesions and the jaw drops open. Nuclear disease is rare, but may occur in multiple sclerosis, syringomyelia and pontine hemorrhage. In this condition atrophy takes place, the normal jaw-jerk is not present and reaction of degeneration occurs. In one of the writer's cases a bilateral masseter paralysis took place in the course of Landry's paralysis after the condition had traveled up the cord. The patient had an infected gall-bladder, which was probably the cause of the Landry's paralysis. She finally recovered from all her paralyses, the masseter difficulty clearing up last.

FACIAL NERVE

The facial nerve is a motor nerve for the muscles of the face performing expression. As we shall consider the intermediate nerve (pars intermedia, nerve of Wrisberg) a separate cranial nerve, we shall not, of course, consider it the special sense portion of the facial nerve as has been done. In the usual descriptions the corda tympani, supplying taste fibers, is considered a part of the facial nerve, but, in our opinion, it is more justly regarded as the peripheral portion of the intermediate nerve and will be here so considered and described.

The facial is the motor nerve of all of the muscles of expression in the face, of the platysma and buccinator, of the muscles of the external ear and of the posterior belly of the digastric and the stylohyoid muscles. The nerve arises in a series of nuclei in the floor of the fourth ventricle and in the pons. The fibers from these nuclei hook around the nucleus of the abducens nerve and emerge from the posterior border of the pons

and go on through the cerebellopontine angle close to the trigeminal and the cochlear nerves. The nerve passes forward and outward upon the middle peduncle of the cerebellum, enters the internal auditory meatus, at the bottom of which it enters the fallopian aqueduct in the petrous portion of the temporal bone, and makes its exit from the skull through the stylomastoid foramen. At the stylomastoid foramen it divides into three large branches to supply, speaking roughly, the muscles of expression of the upper, middle and lower face, together with the muscles of the ear, the posterior belly of the digastric and the stylohyoid.

The facial nerve is probably more frequently paralyzed than any other nerve in the body, and its peripheral paralysis will be described under the caption of *facial palsy*, or *Bell's palsy*. The nerve is variously paralyzed in accordance with the site of the lesion along its course.

A *cortical, or capsular, lesion* most usually affects the lower distribution of the nerve. In this paralysis, the lower face is drawn to the sound side, the angle of the mouth sags and the glossolabial fold is lessened on the affected side. The eyes can usually be closed and the forehead wrinkled. In *pontine lesions* the palpebral fissure may be widened or narrowed and the outer angle of the eyebrow is lowered. The nerve is usually affected in *cerebellopontine tremors*. In *paralysis agitans* the loss of facial expression is bilateral and due to muscular inelasticity rather than facial nerve palsy. The facial nerve may be affected in certain of its muscles in *poliocnecephalitis*, when the affected muscles atrophy. Taste is not affected in nuclear lesions. This fact is one of the arguments that might be used in not considering the chorda tympani a branch of the facial. The facial nerve is often paralyzed in *fracture of the base* of the skull. Bilateral facial palsy is rare but may occur in post-diphtheritic paralysis, basal meningitis (especially syphilitic), encephalitis, and double otitis media. The facial nerve is at times severed during operations on the mastoid, and, when this has occurred, movement may be regained after surgical anastomosis with a cervical nerve or the hypoglossal nerve of the same side has been performed.

Spasms and Tics.—The facial nerve is subject to disturbance, either unilateral or bilateral, in various tics, habit spasms, and from chorea. Meige, White, and others point out that certain of the spasms and tics of the facial nerve are purely psychogenic in origin. Meige distinguishes spasms which originate in emotions and then become habitual, and to these E. W. Taylor has added reflex spasms,—the so-called tic convulsif—as psychogenic. Taylor^a further says, “As distinguished from this type the true facial spasm is not dependent upon the mental state, but is due rather to material changes in the reflex arc either in the sensory trigeminus, facial nucleus or facial stem. . . . This type of spasm,” he says, “usually occurs in the whole distribution, or in definite portions of the nerve, and is usually one-sided and presumably is not influenced by suggestive measures.” It might be added that in the psychogenic cases there is always a constitutional neurotic make-up.

Treatment.—Boissand and Raymond in Paris, Patrick and others in this country, have injected **alcohol**—40 to 70 per cent.—into the facial

nerve. Paralysis takes place and in time complete restoration may recur, often minus the tic. Relief has also been obtained by **surgically stretching the nerve**. In the psychogenic cases, **personal isolation, removal from the usual surroundings, massage, and persuasion**, have produced good results. In either type of case, Meige has suggested a method of **educational control** that the author has found extremely useful. Briefly stated, the patient is made to **stand before a mirror** once or twice daily and at first try to control his facial movements for a few seconds only. This time is very gradually increased to ten or more minutes, after which the patient, not infrequently, gains permanent voluntary control over the muscular movements. This treatment is performed under a competent director who times the patient and the increase should not be more rapid at first than one or two seconds at a treatment.

Meige and Feindel showed that a prototype of a sufferer from tic is a person with a neurotic heredity. They draw a rather involved differentiation between a tic and a spasm as far as their mechanism is concerned, but Boissand makes a definite clinical distinction when he says, "Spasm knows no control, . . . as regards tic, however, inhibition is possible because the phenomenon is cortical. In almost every case reinforcement of the will can momentarily at least check it." In the psychogenic cases of tics other than facial, the author has also found the same educational treatment, especially when combined with the so-called psychic effect of the use of faradism or the actual cautery, most useful. Three of these cases had violent tics of the abdominal muscles only. All three of these were in young women, and psychogenic in origin, the movements more or less resembling coitus, and all yielded to educational voluntary control, beginning with a few seconds and increasing, plus cauterization of the back with the actual cautery. In giving this treatment for tic one must not neglect general hygienic measures and the correction of any pathological condition.

The question of spasm and what is termed "habit-spasm" is an interesting story. When Boissand says "spasm knows no control" he must have had in mind only organic spasm from nerve tract irritation when inhibition was interfered with and not habit-spasm which, without doubt, is practically indistinguishable from tic. Habit-spasms may occur from mimicry or the repetition of a movement to counteract some discomfort as, for instance, stretching the platysma when the collar is uncomfortable, or twitching the nose when there is an abrasion in the nostril. Each time the muscular movement is performed the impulse passes more automatically until it is kept up when there is no one to mimic, or the abrasion in the nose is long since healed. We must remember that all habits, good and bad, have one underlying basis; i.e., a desire on the part of the individual to better his condition. If he were perfectly satisfied, he would contract no habit to obtund some sorrow, to make himself feel physically better, to give himself some supposed sort of satisfaction;—his judgment may be in error, but his desire is to better his condition. For the same reason habit-spasms are formed and are kept up, first because repetition of impulse makes it easy, and second because the impulse becomes auto-

matic and beyond the inhibition of the patient, unless that inhibition is reinforced either by a strong use of the patient's will power or by the aid of the will and direction of another. Another factor which occasionally comes into the control of a habit-spasm or tic is an emotional shock or an illness great enough to make the patient forget the movement. The author remembers seeing a severe case of facial tic disappear during the course of a severe case of scarlet fever.

To educate a patient to control a tic the supervision of a physician, a nurse, or an intelligent member of the family is necessary and the procedure must not be left solely to the patient.

Certain medicines seem to have a beneficial influence in these cases, especially **arsenic**. It is very important that every focus of physical and mental irritation should be removed. Special care should be taken to see that no eye, ear, throat or nose trouble is neglected. It is also important that the physician gets the full coöperation of the patient.

Facial Nerve Paralysis: Bell's Palsy.—Bell's palsy, or peripheral paralysis of the facial nerve, may be due to a lesion anywhere below the geniculate ganglion. A lesion high up in the Fallopian canal by extension from injury to the geniculate ganglion may give, in addition to the facial paralysis, herpes in the auricle and external auditory canal. A lesion in the canal also gives loss of taste to the anterior two-thirds of the tongue on the affected side by its interference with the taste fibers through the corda tympani extension of the intermediate nerve which are in close proximity. A lesion at the stylomastoid foramen causes paralysis of the corresponding side of the face without change in taste.

Etiology.—Bell's palsy may be due to a number of causes. Sleeping in a draft, placing the side of the face against a cold substance, as a window pane, or other exposure, may be the cause. It may follow focal infection, especially pharyngitis, otitis and mastoiditis. It may also be due to syphilis. It may be also caused by trauma from pressure, or blows or operation, on or about the mastoid or the ear. It has occurred after Gasserian ganglion operation.

Symptomatology.—The condition usually appears in early adult or middle life. It starts with brief twitching and slight distortion of the affected side of the face followed by loss of motion, usually in all three branches of the nerve. There may be pain in the ear or taste may be affected, according to how high up in the Fallopian aqueduct the inflammation may extend. The patient can not whistle or puff out the affected side of the face. The brow, because of paralysis of the occipitofrontalis, can not be elevated or vertically wrinkled on the affected side, nor can the eye be completely closed except at times during sleep. The mouth is drawn to the sound side and the tongue, on protrusion, appears to go to this side. The labionasal line disappears and the face looks flabby and expressionless on the paralyzed side. Reaction of degeneration may be modified or complete, and atrophy may take place. The affected eye waters and saliva often dribbles from the corner of the mouth. Hearing may be affected from cochlear involvement if the lesion is high enough up. The patient frequently complains of a sense of stiffness

and discomfort over the paralyzed area. Deep sensibility is not impaired.

Treatment.—Recognizing that rapid restoration of function depended upon allaying the inflammation and reducing the swelling of the nerve at the earliest possible moment, several years ago the author began to apply, in all cases, a **fly blister** over the mastoid and stylomastoid foramen and at the same time, for several days, to apply an **ichthyol** or **antiphlogistin poultice** over the affected side of the face. This procedure obtained the most gratifying results, especially in those cases seen the first few days of the paralysis.

Sodium salicylate, potassium iodid and strychnin are useful in the treatment of Bell's palsy and all may be given at the same time in moderate doses. They should be begun as soon as the case comes under observation.

Massage is of great value, especially if given by a skilled manipulator, and should be applied once or twice daily to both sides of the face. If given on both sides, it not only has the effect of aiding in the restoration of function to the paralyzed side, but prevents permanent contraction of the sound side. The massage movements on the affected side should be from the periphery toward the tip of the mastoid.

Faradic electricity given for from 5 to 7 minutes, once or twice daily, with the electrode frequently lifted and reapplied so as to get clonic, as well as tonic, contraction, is of service. **Bathing the face in alternate hot and cold water** a few moments at a time is also useful. In syphilitic cases, **antisyphilitic treatment** should be employed. Most syphilitic cases are central rather than peripheral.

If the condition does not recover, **anastomosis** with one of the cervical nerves or the hypoglossal may give satisfactory results. In traumatic cases, where the nerve is severed, function may be restored by **uniting the proximal and distal ends of the nerve**.

Prognosis.—The prognosis is good for complete recovery in most cases. Normal restoration of movement usually takes place in from three or four weeks to eighteen months. Some cases never recover entirely, but it is rare that there is not considerable improvement. The more complete the paralysis is, and the higher up in the canal it occurs, the more delayed are the signs of improvement, the more profound is the reaction of degeneration and the worse is the prognosis. In some long-continued cases the muscles on the sound side, from prolonged retraction due to the fact that the antagonistic muscles are not functioning, become contracted to such an extent that even when movement on the affected side is restored they continue to overact and the muscles of the face remain asymmetrical. Although a few cases recover spontaneously, the longer treatment is delayed the more guarded should be the prognosis.

Pathology.—The pathology, in most non-traumatic cases, is that of congestion and swelling of the nerve, which may stop at the stylomastoid foramen or be continued up to the fallopian aqueduct; or the inflammation may begin in the canal and extend downward.

Hunt's Syndrome.—There is a syndrome, described by James Ramsey Hunt, due to disturbance of the geniculate ganglion, in which there is herpes of the auricle and external auditory canal, an extension of the inflammation of which gives facial palsy. Tinnitus and difficulty of hearing occur, and, at times, dizziness, nystagmus, and nausea and vomiting. Sometimes there is otalgia without herpes. This syndrome is fully described under the Intermediate Nerve.

Central Lesions.—Lesions of the facial nerve above the geniculate ganglion, due to tumor, meningitis or syphilomeningitis, may occur. Lesions back to the pons nearly always involve other cranial nerves, especially the trigeminal and vestibular, but also the abducens and hypoglossal. Vomiting, headache and choked disk may or may not be present. Taste is unimpaired.

Lesions within the substance of the pons usually give facial paralysis without affecting taste or hearing, but are accompanied by abducens paralysis from interference of the abducens nucleus in the pons.

INTERMEDIATE NERVE

The intermediate nerve, or nerve of Wrisberg, as it is frequently termed, is a mixed nerve and has been considered the special sense portion of the facial nerve, and, again, as the "thirteenth nerve." The intermediate nerve is a mixed nerve, for it has afferent taste fibers and efferent excitoglandular (vegetative) fibers. The intermediate nerve apparently arises in the medulla oblongata¹⁰ and joins the facial nerve in the internal auditory meatus. The same authority, Spitzka, says that a portion of the intermediate nerve is efferent (excitoglandular) and arises from the nucleus salivatorius. The central portion of the ganglion-cells end in the upper end of the nucleus of the glossopharyngeal nerve. The trophic center of the intermediate nerve is in the cells of the geniculate ganglion.

The intermediate nerve gets its name from its position between the facial and the cochlear nerves at the base of the skull. From the geniculate ganglion some fibers accompany the great and small superficial petrosal nerves, while others accompany the facial and form the corda tympani. The intermediate nerve carries taste impulses to the brain and also contains autonomic efferent fibers which join the submaxillary ganglion.

In the fallopian aqueduct, the intermediate and facial nerves run together for a short distance, but the fibers of the intermediate turn upward and forward in a distinct canal and enter the tympanic cavity through an aperture and leave it through the canal of Hughier. From thence it goes down on the inner side of the spine of the sphenoid, which it may groove, and joins the lingual branch of the trigeminal nerve and supplies taste to the anterior two-thirds of the tongue. Hence the corda tympani extension, according to our conception, is not a branch of the facial nerve, as it is usually described, but is a continuation of the intermediate nerve. It has fibers which seem to give it a ganglionic con-

nection, as far as taste is concerned, with the trigeminal nerve from the geniculate to the gasserian ganglion, and with the glossopharyngeal through the petrous ganglion.

The chief function of the intermediate nerve is to supply, through the corda tympani nerve, taste to the anterior two-thirds of the tongue. It also has trophic, autonomic, sensory and secretory excitor functions. The corda tympani may be considered the peripheral extension of the intermediate nerve from the ganglion.

Hunt's Syndrome.—In 1907 and 1908, James Ramsey Hunt described a herpetic inflammation of the geniculate ganglion which has since become known as Hunt's syndrome. Hunt considered the intermediate nerve as the sensory part of the facial nerve, having its origin in the geniculate ganglion. We, of course, do not take this view but consider the intermediate as a separate cranial nerve as just described, but this does not interfere with the clinical features of the syndrome.

Hunt described a zoster zone for the geniculate ganglion in the interior of the auricle and the external auditory canal, which had been previously thought to belong to the trigeminal distribution and due to gasserian ganglion irritation. He divided his syndrome into three types: (1) uncomplicated auricular herpes; (2) auricular herpes with facial paralysis; (3) auricular herpes with facial palsy and auditory symptoms. The last two types are due to the fact that the intermediate nerve lies between the facial and the cochlear and vestibular nerves and inflammation of the intermediate may implicate, of course, either the facial, giving palsy, or the cochlear, producing deafness or tinnitus, or at times the vestibular, causing vertigo.

The symptoms of Hunt's syndrome are slight febrile disturbances followed by otalgia of a neuralgic character, followed in turn in a few days by herpes of the tympanic membrane.

COCHLEAR NERVE

The cochlear nerve has been described as one of the branches of what used to be termed the eighth or auditory nerve. However, Jelliffe and White in their text-book published in 1917 very properly state: "The eighth cranial nerve is in reality two separate nerves, with distinctly different structures, pathways and functions. It is not a single nerve with two parts. The two nerves are the cochlear or auditory proper, and the vestibular—a portion of the cerebellar apparatus." They also go on to say, "The former handles sounds, the latter serves to orient the body in space. Their receptors lie closely related in the sphenoidal bone! By reason of this close topographical relationship, infections of the middle ear are apt to involve both structures, and by reason of the close associations with cranial structures, brain involvements such as meningitis, abscess, etc., may result. Their central stations are wide apart in the temporal cortex and cerebellum respectively."

The cochlear nerve is the nerve of sound and the receptors of sound

stimuli lie in the organs of Corti in the cochlea. From here the fibers gather and pass close to the facial nerve through the auditory canal, then on through the pontocerebellar angle to enter the medulla, and are relayed in the tuberculum before they go on to the auditory centers in the temporal lobe.

The function of hearing may be tested roughly by closing one ear with the finger and bringing a watch from a distance toward the other ear, noting where the tick is first heard. This may be controlled by comparison with an ear of normal hearing. Sound application, when tested in this manner, is known as aërial conduction of sound, and the deafness may be due to middle ear disease or to nerve deafness. Finer tests should be made with a tuning fork for the recognition of various tones by their number of vibrations.

Nerve Deafness.—Nerve deafness is a condition which may occur from lesion of the cochlear nerve and is a term used when the bone conduction of sound is poor or absent by various tests. For these tests, a watch or tuning fork is pressed over the mastoid bone and, if the sound is heard normally, the nerve is said to be intact and bone conduction good. If the instrument is placed against the center of the forehead, and the sound is heard well in one ear and not in the other, bone conduction is said to be poor on the side of the deficient appreciation, and this indicates a lesion of the cochlear nerve. Before making these tests it must be determined that the ear drum is not ruptured and that there is no wax occluding sound to the ear. In testing one ear, the other ear should be closed to sound by the operator's finger. In some cases air conduction of sound is absent from disease, but if tested for bone conduction the nerve will be found intact. On the other hand, there are conditions in which bone conduction appreciating sound through the nerve is absent, but sound can be heard through air conduction. The sound-conducting apparatus may be so diseased that sound cannot be heard either through air or bone conduction. The gutta-percha fan held between the teeth by certain deaf people is used to bring out the sound through bone conduction.

Reinné noted that in disease of the labyrinth there was a diminution of both aërial and bone conduction of sound, the bone conduction being frequently lost, while in disease of the middle ear the bone conduction is increased and air conduction is diminished. For other tests and refinements of the above, the reader is referred to books on otology. A lesion of the cochlear nerve may cause nerve deafness or it may cause tinnitus.

Tinnitus.—Tinnitus is a condition which includes various subjective sounds and noises in the ears. A roaring sound such as is heard when a seashell is placed to the ear is probably the most common, or such phenomena as ringing, singing, buzzing, ticking, hissing and whistling may occur. In hyperemic conditions these sounds are increased when the patient lies down and in anemic conditions when the head is elevated. Certain drugs, notably quinin and the salicylates, produce the sounds. Tinnitus may also occur after the ears are boxed or when there

is water, wax, or other substance in the external auditory canal. Disease of the labyrinth and obstruction to the eustachian tube will also cause tinnitus. It is common in chronic, middle, or internal ear disease.

Throbbing may be annoying in the ear in hyperemic cases. In certain cases this may be controlled by pressure on the carotid artery. In hysteria and migraine various subjective sounds may occur. Often this is a sense of crackling, deep in the head, or a feeling that blood is rushing through a certain area of the brain.

Monotonous noises, as a ticking clock, or the hammer sounds of a boiler shop, may persist long after the sounds have ceased, but this can not be considered true tinnitus.

Treatment.—Tinnitus may be lessened by the administration of **bromids and belladonna, potassium iodid, galvanism**, and the application of a **fly blister** behind the ear. The injection of **pilocarpin** one minim with a small hypodermic needle into the middle ear helps in some cases. In cases of cerebral hypertension, **purges veratrum viride or nitroglycerin** help, and in anemic cases **iron, arsenic and massage** are of value.

Variations in Acuity of Hearing.—The sense of sound, as does the sense of smell, varies greatly in intensity. The American Indian puts his ear to the ground and hears sounds we cannot detect; the trained musical ear hears tones not detected by others; the trained chest examiner hears murmurs and râles the student cannot appreciate. On the other hand, some have a congenital deficiency of hearing. Some sounds, especially monotonous sounds, may be voluntarily or involuntarily ignored. The tick of a clock in a room is usually only heard when attention is attracted to it and can frequently be ignored when it becomes annoying. Workers in boiler factories and machine shops do not hear the noises because they become accustomed to them. In talking amid these noises they rarely raise their voices, but if an unusual sound occurs in the shop their attention is immediately attracted to it.

Hallucinations of Sound.—Hallucinations of sound are in a sense cortical in origin and are usually due to the misinterpretation of some ordinary sound. A creaking shutter may be interpreted as a voice, or the wind on telegraph wires as singing. In fact, it is not hard for a normal person to listen to a grandfather's clock and hear it say words to him.

Mechanism of Hearing.—The projection of sound is not entirely understood. Singing, music, voices, noises surround us which we do not hear for our ear is not so attuned. But when a radio properly set is placed by us these sounds, though originating thousands of miles away, immediately become audible. Sound travels in waves through the ether of the air and these waves consist of molecules of air in longitudinal vibration. These waves are but little disturbed by surrounding vibrations of the air, unless they are of the same periodicity. These sound waves, or vibrations, enter the ear and strike the tympanic membrane and set it into the same kind of vibration; then the ear bones and fenestra set the perilymph into similar vibrations; this affects the cochlear membrane and the sound becomes appreciated through the rods of the organ of Corti in the cochlear. The cells of the cochlear are bipolar, one fiber

arborizing around the rod of the organ of Corti, and one going toward the brain in the cochlear nerve. Combinations of the vibrations, or notes, cause sensations of harmony and discord, and appreciation of these notes in individuals may differ both quantitatively and qualitatively.

VESTIBULAR NERVE

The vestibular nerve was formerly described as a part of the old eighth or auditory nerve. For reasons for considering the vestibular nerve as a separate nerve, the reader is referred to the discussion under the heading Cochlear Nerve. The vestibular nerve is the nerve of equilibrium, through the appreciation of gravity, and conducts impulses from its receptors in the semicircular canals, utricle and saccule, to the cerebellum.

The vestibular nerve may be considered to arise in the bipolar cells of the vestibular, or Scarpa's ganglion at the bottom of the internal auditory meatus. Its peripheral fibers go to the special sense organs in the semicircular canals, the utricle and saccule, and its central fibers chiefly form the auditory nucleus in the floor of the fourth ventricle, from which fibers are sent to the cerebellum, with those of the lateral columns of the cord and of the ocular nuclei. Owing to these connections, lesions of the nerve may be manifested by nystagmus, incoördination of the head, neck and body and, chiefly, by vertigo.

Vertigo is common in cerebral tumor but more pronounced in tumor of the cerebellum. It may also be a symptom of nervousness, disease of the middle or internal auditory canal, cerebral arteriosclerosis, approaching syncope, migraine, epilepsy, stomach and intestinal disorders, other toxemias and various conditions to be briefly discussed. However, it is believed that in all vertigoes the vestibular apparatus is involved.

Sense of Equilibrium.—Equilibrium is that sense which serves us to orient ourselves in space and is concerned with sensations, through the vestibular nerve, from the semi-circular canals and vestibule which have a peculiar influence upon complex muscular activity. The semi-circular canals lie in three planes, nearly at right angles to each other, and contain fluid. The special sense vestibular nerve-cells are affected by the pressure of the endolymph in the semi-circular canals and the nerve impulses, engendered reflexly, affect the tone and coördination of muscles in conjunction with other cerebellar impulses, giving synergy, muscular, visual and tactile faculties. This complex is still further involved by the fact that it seems certain that the cells of the utricle and saccule have an accessory function in equilibrium, probably as Breuer suggested, serving to give us information, when the head is at rest or when it is progressing forward or backward in a non-rotary motion, as to its position. The rotary movements are told through the semi-circular canals. Members of the aviation service have remarkably little difficulty with loss of equilibrium. This is probably due to many factors, prominent among which must be the fact that their bodies are more or less fixed in the seat of the machine, and hence receive a sense of stability and support, and, also, that the rotations the airplane performs are of wide excursion

and the displacement of the fluid in the semi-circular canals is less sudden than, for instance, in whirling in a revolving chair. Apropos of the sense of equilibrial satisfaction one feels if fixed in position when whirling in the air, the writer well remembers the sense of stability and lessening of the sense of vertigo he experienced while riding in the whirling "boats," swung high in the air, at the St. Louis Exposition. At first he did not strap himself to the seat and experienced a most disagreeable sensation as if he was about to fly off into space, which was greatly minimized when he adjusted the seat strap around his body.

Ménière's Disease.—In 1861 Ménière described the labyrinthine vertigo which bears his name. Other vertigoes may be differentiated as they affect different parts of the vestibular nerve. For instance, the first neuron may be affected, or the nuclei in the medulla, or the lesion may be in the region of the posterior longitudinal bundle, or there may be a lesion of the visual tracts implicating the vestibular nerve, or the chief trouble may be in the cerebellum.

Etiology.—Many causes have been suggested for the condition. Hemorrhage into the labyrinth occurring in the course of syphilis, leukemia or arteriosclerosis may be the cause, or extension of the perilymph of angioneurotic character, or the beginning of a vestibular tumor may be manifested by symptoms of Ménière's syndrome.

Symptomatology.—Ménière's disease, or labyrinthine vertigo, is often accompanied by nystagmus. Jelliffe and White state: "Should the nystagmus last for twenty-four hours, or more, it is of intracranial origin. If it lasts a shorter interval and is uninterrupted by quiet intervals it may be peripheral or central." The disease may be either *acute*, which is of short duration and a rare form, or *chronic*, which is more or less common. The condition is marked by exacerbations, or attacks, with freedom, or comparative freedom, between the attacks. In the intervals between the attacks, the Bárány caloric reaction is diminished on the affected side. In the milder forms of Ménière's disease there may or may not be tinnitus or buzzing on the affected side and hearing is not impaired. In the severer forms, there may be both deafness and tinnitus, with giddiness and reeling, and nausea and vomiting. The vertigo may, or may not, come on from change of position and may be so sudden as to throw the patient down. There may be momentary loss of consciousness, but this is not the rule. Nausea, vomiting and headache follow the vertigo. Vertigo may be unilateral or bilateral, but the tinnitus is usually unilateral. Often this group of symptoms is attributed to a disordered digestion, the nausea and vomiting attracting more attention than the vertigo.

Diagnosis.—The disease has been mistaken for epilepsy but a careful history of the attacks will show the difference.

Cerebellar vertigo should not be confused with Ménière's syndrome; for, in the former, the gait is unsteady, with the eyes open or closed, and symptoms are usually absent and there is adiadokokinesia and asynergy.

Treatment of Ménière's Disease.—The treatment of Ménière's disease is first to apply a fly blister, back of the ear. This alone is frequently of much benefit. Babinski recommends repeated lumbar puncture. In my

experience the most reliable remedies, besides the blister, have been **belladonna, bromids, suprarenal gland extract, and potassium iodid.** The treatment should also include considerable **rest in bed and massage.** Every case should be examined by a competent aurist. A Wassermann reaction should be made in each case, and if positive, **salvarsan and other antisyphilitic remedies** should be pushed. **Nitroglycerin and sweating** are useful if there is circulatory hypertension. Frequently the repeated injection of **one minim of pilocarpin** through the drum with a small hypodermic needle into the labyrinth every few days is of great value; this should always be tried in obstinate cases. Of course, it should not be attempted by any other than a competent aurist.

Prognosis.—The prognosis is uncertain. Many cases, however, recover. Huthcheson¹¹ states that the vertigo often decreases as deafness becomes more complete.

Bonnier's Syndrome.—Bonnier's syndrome, due to interference with Deiter's nucleus, gives the symptoms of Ménière's disease plus those of interference with the glossopharyngeal, vagus and sometimes the trigeminus and oculomotor nerves. The chief additional symptoms are apprehension, tachycardia, hemiplegia, weakness, and at times somnolence. Little can be done for this condition unless it is of syphilitic origin.

Vestibular Vertigos.—Vertigo is a sense of turning or movement, in which the patient feels as if he is turning around or that objects are whirling around about him, and with this there is a slight interference with consciousness.

Vertigo may be a symptom of various brain lesions and the maintenance of equilibrium in vertigo is difficult. Vertigo is often an expression of the aura in epilepsy. Epileptic vertigo can usually be distinguished from vestibular vertigo because, when the epileptic attack is over, the vertigo is not present. Gowers describes an epileptoid vertigo, only so named because it comes in attacks with an entirely free interval between the attacks, in which the onset is sudden. These attacks are induced by fatigue and last for an hour or more. Certain vasomotor changes in the labyrinth may give a vertigo closely resembling true Ménière's disease.

There is another form of vertigo known as *Gerlier's disease*, or paralyzing vertigo, which seems to occur only in Switzerland and there only during the summer months, and affects only those working among cattle, or in the field. The characteristic symptoms are pain in the back of the neck and head, muscular weakness, ptosis, dimness of vision and vertigo. The attacks last only a few minutes.

Vertigo frequently occurs in persons with refractive errors or other eye defects, including ocular muscular imbalance. In almost any cerebellar disease, vertigo is a common symptom and is said to be an especially prominent symptom if the middle peduncle is involved.

Many other conditions may induce vertigo, such as turning rapidly around, looking down from high places, migraine, cerebral arteriosclerosis, and cerebral anemia.

Tests for the sense of balance and for the elicitation of vertigo have

come into especial prominence since the beginning of the world war, because of the necessity of examining men to ascertain their fitness for aerial service. The Bárány test is used in this, and in testing for other forms of vertigo. The general method of application of this test is that of syringing the ear with hot or cold water and putting the patient in a revolving chair which is whirled and then stopped suddenly and making the patient point to some object or perform some other movement. A patient who cannot satisfactorily perform the test, is frequently said to have "vestibular ataxia."

GLOSSOPHARYNGEAL NERVE

The glossopharyngeal nerve is a mixed sensory and motor nerve, giving ordinary sensation to the mucous membrane of the pharynx, fauces and tonsils, and the special sense of taste to the posterior third of the tongue and also some taste-fibers to the soft palate. Its motor fibers supply the middle constrictor of the pharynx and the stylopharyngeus muscle.

The real origin of the sensory fibers of the nerve is in the jugular and petrosal ganglia and their apparent origin in the floor of the fourth ventricle. From the glossopharyngeal nucleus, taste impressions pass to the thalamus of the opposite side and from thence to the gyrus hippocampus, where the cortical taste center is situated. The motor branch is small and arises in the nucleus ambiguus in the floor of the fourth ventricle. The motor fibers pass outward and leave the skull through the jugular foramen external to, and in front of, the vagus and spinal accessory nerves. They then pass beneath the styloid process and the muscles attached to it to the lower border of the stylopharyngeus. They then curve forward and pass beneath the hyoglossus muscle to reach its distribution in the pharynx and mouth.

Paralysis of the glossopharyngeal nerve alone is never seen in man (Purves Stewart), but it is often paralyzed in conjunction with other cranial nerves. When paralyzed there is loss of taste in the posterior third of the tongue, anesthesia of the back of the tongue and upper part of the pharynx, and difficulty of swallowing.

Weisenberg¹² seems to have shown that the lower part of the trigeminal and the upper part of the glossopharyngeal intermingle in their sensory distribution, so that irritation of the glossopharyngeal may give pain closely resembling trifacial neuralgia.

There are filaments from the petrous ganglion of the glossopharyngeal nerve which connect with the vagus and sympathetic nerves. One branch arising from the petrous ganglion is known as the tympanic branch, or Jacobson's nerve, some of whose branches supply tympanic branches to the ear.

Interference with the glossopharyngeal nerve may be told by testing taste on the posterior part of the tongue, or by testing sensation to the pharynx.

To test taste, the patient should be made to protrude the tongue and solutions of salt, bitter, sour or sweet, applied to the portion to be tested on a swab of cotton or camel's hair brush. These substances are the only ones primarily recognized by taste. After each substance is applied, the mouth should be rinsed. Instead of making the patient answer verbally he should indicate the substance by pointing to the word "sweet," "bitter," etc., written on a piece of paper, for in answering verbally the substance may get in the saliva and be distributed to, and appreciated by, other parts of the mucous membrane of the mouth and thus ruin the test. Volatile substances, cologne, alcohol, coffee, tobacco, etc., are not used in this test for they are appreciated more readily by smell than taste.

The glossopharyngeal nerve carries the taste-fibers from the posterior third of the tongue and the palate to the glossopharyngeal nucleus, while the corda tympani extension of the intermediate nerve carries the taste impulses of the anterior two-thirds of the tongue to its nucleus in the medulla, a large portion of them going to the glossopharyngeal nucleus. From here, they go by way of the medial fillet to the optic thalamus of the opposite side, and thence to the gyrus hippocampus, where the cortical taste center is situated posterior to the area of olfaction.

Much discussion has been indulged in as to the course of these taste-fibers from the anterior two-thirds of the tongue, some giving them origin through the trigeminal by a circuitous route, and some think they form a part of the facial nerve. The logical conception seems to be that they belong to the intermediate nerve and are probably relayed in the cells of the geniculate ganglion and in the medulla on their way to their cortical center in the hippocampal gyrus.

It seems to be a fact that the taste appreciation of bitter is more developed on the back part of the tongue and palate than on the front part of the tongue, while on the anterior part and tip of the tongue sweet is best appreciated.

VAGUS NERVE

The vagus nerve, frequently called the pneumogastric nerve, has motor, sensory and vegetative functions and a very wide distribution, including various organs of the throat, thorax and part of those of the abdomen. If such a statement is permissible, it might be said that the vagus is the most important of the cranial nerves.

The vagus nerve supplies the organs of voice and respiration with motion and sensation, and motion to most of the muscles of the soft palate, the muscles of the pharynx, larynx, esophagus, stomach, a good deal of the intestines, the bronchi, and sends fibers to the heart. It gives sensory fibers to the dura, the ear, the pharynx, larynx, trachea, esophagus, bronchi, stomach, pericardium, and is supposed to send fibers to the pancreas, kidneys and suprarenal glands. It gives autonomic fibers, which are motor, for bronchial muscles, sensory for respiratory passages, and inhibitory for the heart. It also gives other autonomic fibers to the stomach and the intestines. Recent authorities include in the vagus what was formerly known as the "bulbar part of the spinal accessory."

which is derived from, and belongs to a continuation of the vagus nucleus (the nucleus ambiguus) in the medulla.¹³

The fibers, which were formerly considered the vagus accessory portion, or, as they were sometimes called, the bulbar portion, are purely motor and arise from large multipolar cells in direct continuation with the nucleus ambiguus. The cells of this nucleus may be considered a part of the vagus nucleus. These fibers in their course give filaments to the other part of the vagus, which are distributed, through the recurrent laryngeal nerve, for the supply of the muscles of the larynx. They also possibly give some filaments to the heart, but the main portion of the fibers supply the azygos uvulae and levator palati muscles.

The origin of all the motor fibers of the nerve for all practical purposes may be considered to be in the nucleus ambiguus in the floor of the fourth ventricle and a continuation of this nucleus. The sensory fibers arise in the ganglion of the root and the ganglion of the trunk of the nerve. The motor and sensory portion join and pass through the jugular foramen, where is situated the superior or jugular ganglion and, after the exit from the foramen, the nerve is connected with the inferior ganglion. It may be that in the future the sensory portion of the vagus will be considered a separate cranial nerve. Its exact relation to the motor portion is not entirely understood, but as they pass in the same sheath, and at times are distributed together, it seems better to treat the vagus, for the present, at any rate, as a mixed nerve.

One branch of the vagus, the *recurrent laryngeal nerve*, should be mentioned. It is the motor nerve of the larynx and has much to do with phonation, as we shall see later on. For a detailed description of its anatomy and that of other laryngeal nerves the reader is referred to the various works on anatomy.

If the lower roots of the vagus are affected by an *intracranial lesion* the hypoglossal nerve is usually also affected. Paralysis of one vagus trunk gives paralysis of the palate and larynx on the affected side, and also anesthesia of the larynx on that side. The motor paralysis may be tested by watching the arch of the soft palate and making the patient say "Ah." The unaffected side of the palate elevates and the uvula is deviated to that side, while the affected side does not elevate but lags.

If both vagi are paralyzed, the heart becomes rapid and arrhythmic, respiration becomes slow and irregular, and gastric pain, dilatation and vomiting may occur. Gastric symptoms may occur in unilateral paralysis also, but cardiac and respiratory symptoms rarely, if ever, occur when one nerve only is affected.

As the vagus distribution is so wide, the symptoms of lesions in various locations may be more or less distinct. The nerve may be disturbed by multiple neuritis, cerebrospinal syphilis, meningitis or by tumor or aneurysm in the cranium. It may also be affected by hemorrhage or softening implicating the medulla, by surgical or other lesions to its trunk or branches, or by disturbance of the vegetative nervous system. The latter will presently be described under Vagotonia.

When the *pharynx* is affected there is difficulty in swallowing, and

if the soft palate is involved, food passes up into the nasopharynx, also in speaking, the patient has a nasal voice. These symptoms occur in bulbar palsy and often in postdiphtheritic paralysis. A spasm of the pharynx is said to occur in hydrophobia, and is also known in functional nervous affections.

When the larynx is affected, especially in recurrent laryngeal paralysis, phonation is interfered with.

Recurrent laryngeal paralysis may occur from compression on the nerve of an aortic aneurysm, enlargement of glands of the neck, mediastinal growths, dilatation of the left auricle from mitral stenosis, or from surgery of the neck or other trauma. In health, the vocal cords lie parallel and almost touch on phonation. They abduct with breathing. If one recurrent laryngeal nerve is paralyzed, the vocal cord on one side becomes fixed midway between abduction and adduction, and, although the patient can speak, his voice is hoarse. In bilateral recurrent laryngeal paralysis, phonation cannot be performed because the vocal cords are rigid and cannot be approximated. It should be remembered that recurrent laryngeal paralysis may occur from lesions of the medulla, or base of the brain, as well as from the nerve itself. The author has seen it in a tumor of the middle fossa affecting the medulla. In this case the pharynx, through the vagus, and the tongue, through the hypoglossal, were both affected.

True Abductor Paralysis.—True abductor paralysis of the vocal cords is never functional. Although the voice is not affected, if it is unilateral, the laryngoscope will reveal that the cord does not move outward during inspiration. In tabes, a laryngeal crisis may occur, and, when it does, the paralysis is abductor in type and is usually unilateral. In bilateral abductor paralysis, the cords can come together but cannot move outward. Hence the patient does not have difficulty in phonation but does have difficulty with respiration and forced expiration is stridulous.

Adductor Paralysis.—On the other hand adductor paralysis is frequently functional. It occurs on both sides and causes the familiar condition known as hysterical aphonia. Reëducation, or faradic shock, will bring the voice back. It is sometimes quite persistent, and the author had one patient in whom it lasted a year or more. The patients, although unable to speak aloud, whisper without difficulty. Hysterical aphonia usually originates in some sexual, or other emotional shock.

Spasm of the Larynx.—Spasm of the larynx may occur, and in this condition the abductors are overpowered by overaction of the adductors. Central or peripheral irritation of the vagus may be the cause of the laryngeal spasm, but the condition is usually hysterical. When the spasm is severe, a hypodermic of morphia, or the inhalation of an amyl nitrate pearl or chloroform, will relieve it.

Anesthesia of the Larynx.—Bilateral, or unilateral, anesthesia of the larynx may be due to lesion of the medulla, diphtheria, injury, or it may be functional. When it occurs, food may enter the larynx without being appreciated and deglutition pneumonia usually results. To pre-

vent pneumonia, the patient should be fed, in bilateral cases, with a **nasal tube** passed well down the esophagus, or in unilateral cases he should be made to **eat slowly and lie on the unaffected side** while doing so.

Hiccough.—Hiccough is due to a spasm of the diaphragm through the phrenic nerves and occurs in exhausting illness, after surgical operations, from a distended abdomen, from hurried eating, occasionally as a reflex in tabetic cases and as a functional manifestation. Hiccough has occurred in epidemic form concurrent with influenzal epidemics. In simple cases drinking water slowly or holding the breath will cause the spasm to cease. In more severe cases faradism to the phrenic nerves or spraying the line of the diaphragm with **ether or ethyl chlorid**, at the same time giving internally a few drops of **chloroform, ether**, or a teaspoonful of **Hoffman's anodyne**, will usually control the spasm. If the abdomen is distended, it should be relieved by an **enema**. In very protracted and severe cases, the author has used with marked success 5 grain (0.324 gram) capsules of **tangue musk**, given at four-hour intervals.

The Vegetative Nervous System.—One of the latest and most important advances in neurology for many years has been the separation of the vegetative nervous system by distinctive description, and the discovery of new facts in the physiology and the symptomatic disturbance of its two subdivisions—the sympathetic nervous system and the autonomic nervous system. Much of the credit for this work is due to Heinrich Higer of Warsaw and to Eppinger and Hess of Vienna. To understand the clinical condition *vagotonia*—soon to be described—we shall first have to get a general idea of the results of the recent investigations of the vegetative nervous system.

The vegetative nervous system is an outgrowth of the cerebrospinal system and on all of its fibers are ganglion cells. The vegetative nervous system is distributed to the non-striated muscles of the body, the pupils, various glands and viscera, the heart, the blood-vessels and the genital organs.

There are certain reflexes connected with the vegetative nervous system, such as salivation, sweating, flushing of the skin and the genital reflexes and also others, as disturbance of certain hollow organs, for instance the stomach, uterus or bladder, through fear, pleasure or surprise. Referred visceral pains, hunger, nausea, the feeling of satisfaction upon emptying the bladder, are experienced through the vegetative nervous system.

The sympathetic system has been best known in the past and its anatomy is more or less familiar. All of the fibers of the vegetative nervous system, not included in what is known as the "extended vagus," compose the sympathetic system. In the discussion of *vagotonia*, we do not deal directly with the sympathetic division, but with the autonomic or extended vagus division of the vegetative nervous system. We must bear in mind, however, that in stimulation of the autonomic system there is more or less compensatory depression of the sympathetic division. The sympathetic and autonomic divisions innervate together and balance control. Purves Stewart likens this antagonistic action to that of a pair

of reins. The sympathetic accelerates the heart and the autonomic inhibits it.

Pharmacological experiments have shown that **adrenalin** is a drug which acts only on the *sympathetic* division of the vegetative nervous system, and it does so by stimulation. A depressant agent for these sympathetic fibers, according to Eppinger and Hess,* is not yet known, although **morphin** does depress their nuclei situated within the central nervous system.^{14 15} It has also been found that **pilocarpin** and certain other drugs stimulate the *autonomic* system, while **atropin** depresses it.

Vagotonia.—"Vagotonia is a lasting tonic irritation of the vagal part of the autonomic system which maintains its end organs in a state which very closely resembles that produced by electrical stimulation of the autonomic," say Eppinger and Hess, and it is thought by them, and others, that this tone irritation is due to, and kept up by, the under secretion or oversecretion of one or more of the ductless glands. This field offers splendid opportunities for future investigation.

Vagotonia includes much that was previously relegated to neurasthenia, hysteria, anxiety, fear states and general nervousness, and vagotonia practically takes the place of what were known as the visceral neuroses and the so-called vasomotor neuroses. Vagotonia accounts, in part, at least, for many asthmatic conditions, pseudo-angina, vasomotor gastric hyperacidity, pylorospasm, and has a definite relation to exophthalmic goiter, the nausea and vomiting of pregnancy, certain states of diarrhea, especially mucous colitis, and to spastic constipation. Vagotonia also embraces some hitherto unexplained cases of bradycardia, arrhythmia, vague gastric distress and aërophagia. Vagotonia with *tachycardia* is often associated with visceroptosis, and it should be remembered that vagotonia may be associated with other diseases, either organic or functional.

Symptomatology.—Vagotonia is a functional condition, the symptoms of which are numerous and may be major or subsidiary. Part of the extended vagus may be affected, or practically the whole of the autonomic system at the same time.

An individual may be more or less uncomplaining, but still be said to belong to the vagotonic type, when he has gastric hyperacidity, eosinophilia, bradycardia, slight arrhythmia, somewhat spastic constipation and sweaty hands and feet. Persons of this type are said to be highly susceptible, especially with regard to salivation and sweating, to even small doses of pilocarpin.

* This statement by Eppinger and Hess may be disputed by following references applying to apocodein and nicotin:

BASTEDO: Textbook of Materia Medica, Pharmacology and Therapeutics, 1914, page 104:

"It (apocodein) is employed in the laboratory as a general paralyzant of sympathetic nerve endings. In this respect it is directly antagonistic of epinephrin. . . . It acts by cutting off splanchnic control of intestinal activities through the depression of the sympathetic nerve endings."

CUSHNY: Textbook of Pharmacology and Therapeutics, 6th ed., page 435:

" . . . resembles *nicotin* in paralyzing the sympathetic ganglia."

Besides these symptoms vagotonics may have irregular breathing, occasional tachycardia when associated with visceroptosis or hyperthyroidism, nervous and anxiety states, localized hyperhidrosis, flushings, nausea and other gastric distress, cool damp hands, acne, and intestinal symptoms, as spastic constipation, alternating constipation and diarrhea, or mucous colitis. These patients are usually constitutionally inferior. They often have enlarged tonsils and adenoids, husky voices, tremors of the hands, tongue and eyelids, and lessened sensibility to pharyngeal irritation. They urinate frequently and have an excess of phosphates and oxalic acid. They are startled by noises, easily frightened, apprehensive, often peevish or self-accusatory, sexually hyperexcitable and usually poor sleepers.

At times some or all of these symptoms undergo an acute exacerbation and we observe a nervous "spell" or "attack," the description of which by the patient is usually vague. Gowers, years ago, described some of these "spells" under the name vagal attacks. The attack consists chiefly of a marked increase in the apprehension and fear symptoms, sometimes to a sense of impending death, rigors or "nervous chills," prostration, acute functional cardiac, respiratory, and gastric distress, often pains resembling visceral crises and marked sweating.

Treatment.—Fortunately for these acute attacks, as well as for the chronic states, we know a remedy which is specific—**atropin**. This drug depresses the overstimulated autonomic system and may be given in either tablet form or in solution. It may be administered under the skin or by the mouth. For chronic cases the author prefers it in solution, by mouth, about 1/120 grain (0.00054 gram) of **atropin sulphate** three times a day. For the acute exacerbations 1/100 grain (0.00065 gram) may be given by hypodermic or on the tongue.

SPINAL ACCESSORY NERVE

The spinal accessory nerve is a motor nerve, and what was formerly considered the accessory part of the vagus is now included in the vagus system.

The spinal accessory nerve is entirely motor and has its origin in cells in the cervical region of the cord, in the gray substance of the intermediolateral tract. The roots from this origin form a trunk which ascends and enters the skull through the foramen magnum and then turns outward to leave the cranial cavity through the jugular foramen where it receives filaments from the vagus. The nerve passes under the digastric and stylohyoid muscles, pierces the sternomastoid muscle to which it gives fibers, and passes on to terminate in the trapezius muscle.

A destructive lesion of the spinal accessory nerve gives partial paralysis of the sternomastoid and trapezius muscles. This paralysis is not complete, however, because these muscles receive other motor fibers from the cervical plexus.

One way of testing for a lesion of the spinal accessory nerve is to get the patient to attempt to shrug the shoulder on the affected side. When there is a lesion of the nerve, the affected shoulder is shrugged with difficulty, or not at all.

Irregular spasmodic action of the sternomastoid and trapezius muscles is seen in syphilis of the nervous system, multiple sclerosis, some cases of epilepsy and other brain conditions. Nuclear affections are rare. Peripheral palsies cause reaction of degeneration, atrophy and loss of reflexes. When the trapezius is paralyzed there is an altered neck line, and displacement downward and rotation outward of the scapula. Tics or wry-neck may be psychogenic or cortical in origin.

Torticollis.—Torticollis, or wry-neck, may be fixed or congenital, labyrinthine, professional, neuralgic, or spasmodic.

Congenital or fixed torticollis is due to unilateral contraction of the sternomastoid from birth trauma. The head inclines to one side and can not be moved to the other side. There is a fixation of the muscle rather than spasm.

Labyrinthine torticollis is a voluntary inclination of the head to the opposite side, in order to lessen the vertigo, in chronic irritation of the semicircular canals.

Professional torticollis is an occupation neurosis occurring in certain professions, as shoemaking and tailoring, when the head is turned frequently to one side.

Neuralgic torticollis occurs during attacks of neuralgia and ceases when the neuralgia disappears. It is spasmodic and tonic in type.

Spasmodic torticollis is a severe form of tic or habit-spasm occurring in the muscles supplied by the spinal accessory nerve. It may be tonic, irregularly clonic, or combined tonic and clonic in character. For some unknown reason the head is usually drawn to the left side and it is sometimes jerked backward or, as in one of the writer's cases, forward. Sometimes there is a combination of backward and lateral or forward and lateral jerking. The muscles on both sides are generally increased in tone and hypertrophic. The movement ceases during sleep. It usually begins by spasmodic contractions, but later on the position of the head may become more or less fixed.

Most cases are psychogenic in origin, but some appear to be due to either a reflex or toxic condition which may be traced to some infected foci. When it is psychogenic in origin it occurs in persons of a psychoneurotic makeup. The condition is rare in childhood and usually occurs during or near middle life.

Treatment.—The treatment of torticollis depends upon the type and causation. In *congenital torticollis*, the contracted muscles may be lengthened by surgery. Other than surgery, treatment is of little benefit in the congenital form. In *labyrinthine torticollis* local aural treatment may be of assistance, or the general measures used in the treatment of aural vertigo may be tried. *Professional torticollis* is treated by a change of posture, or of vocation, together with massage and active and passive movements to the neck. For *neuralgic torticollis* the gen-

eral treatment of neuralgia with **anodynes** and **analgesics** in conjunction with **massage** and the application of **dry heat** is effective.

The treatment of *spasmodic torticollis* is much more complicated. The case should be considered as possibly of toxic origin, of psychic origin, or of both toxic and psychic origin. In toxic cases, or, for that matter, in any case, **local infections** in the teeth, throat and sinuses should be **searched for and removed**. It is in these cases that the **salicylates** given in frequently repeated doses have a good effect. **Gaultheria liniments** are also useful. **Massage, hydrotherapy** to the back of the neck, and the application of **dry heat** by "ironing," or the use of a **leukodescent lamp**, are beneficial.

As there is present, in most cases of spasmodic torticollis, a large psychic element, a **careful history should be taken**, including the noting of family peculiarities, investigation into the habits and a search, by psycho-analysis, for some emotional or sexual factor in the genesis of the condition. When such a factor can be found **psychotherapy** by suggestion and reëducation should be tried. In spasmodic cases, forcible movement and extension of the neck, massage, hot douches to the neck and shoulders, the use of the actual cautery, or the application of dry heat, are beneficial. If possible these cases should be treated away from their abodes in a sanatorium or properly equipped clinic.

Sedatives, as the **bromids**, small doses of **tincture of opium**, **cannabis indica**, or **hyoscyamus** may be required. In some cases **mechanical support** by a correctly adjusted collar of leather or plaster-of-paris is needed or, for a while, the patient may be put to bed with **extension by weights** applied to the head. In persistent cases **division of the spinal accessory nerve** is necessary, or the **lengthening of the muscle by surgery**.

HYPOGLOSSAL NERVE

The hypoglossal nerve is a motor nerve and supplies the entire motion of the tongue. It has its origin in the floor of the fourth ventricle. Its fibers pass outward and leave the skull through the anterior condyloid foramen. After its exit from the skull it is joined by branches from the first and second cervical nerves, which help to depress the hyoid bone, and also by a branch from the cervical sympathetic, which gives it vasomotor function.

A lesion of the hypoglossal nerve gives paralysis, atrophy and wrinkling of the corresponding half of the tongue. On attempt to protrude the tongue the tip turns to the sound side, because the muscles of this side contract and overcome the paralyzed muscles. If the nerve is bilaterally paralyzed the tongue cannot be protruded. As motion of the tongue has a good deal to do with deglutition, swallowing becomes difficult in paralysis of the hypoglossal.

Certain lesions of the hypoglossal nucleus also affect the lowest cells of the facial nucleus and paralysis of the orbicularis occurs as well as of the tongue. These lesions are usually bilateral.

Hughlings Jackson's syndrome consists of hemiatrophy of the tongue and unilateral paralysis of the trapezius, sternomastoid, vocal cord and soft palate, all on the same side. This lesion involves the lowest roots of the vagus and the spinal accessory and hypoglossal nerves.

Tumors, organic brain diseases, or mineral poisons may cause peripheral lesions of the nerve. Poliomyelitis, multiple sclerosis, syphilis or tumors are usual causes of nuclear and supranuclear lesions.



FIG. 3.—EARLY HYPOGLOSSAL HEMIATROPHY.

When the hypoglossal nerve is paralyzed the base of the tongue rises higher on the paralyzed side because of the atony of the hypoglossal muscle. In long-standing cases, fibrillary twitches, especially in nuclear lesions, or tremors, may appear. The paralyzed side shows reaction of degeneration. If both sides are paralyzed, chewing, swallowing and speaking all become difficult. Hysterical paralysis of the tongue has been known. In this condition there is no true atrophy and there are no changes in electrical reactions. The author has noted, in three markedly hysterical cases, elongation and supermotility of the tongue with relaxation of the frenum so that the patients had no difficulty in placing the tip of the tongue in the nasopharynx, or in two cases, curving it over and downward in the throat, semiswallowing it.

Besides hysterical conditions affecting the tongue, other hypoglossal psychogenic disturbances occur, such as ties, lisping, stammering, stuttering and licking and chewing movements.

Mechanism of Speech.—Speech is the expression of human ideas by words, and an elaboration of word combinations forms language which may be both written and spoken. The mechanism of speech is exceedingly complex and involves a considerable cortical and subcortical area of the brain and the function of many of the cranial nerves. Articulation may be interfered with in paralysis of the masseter nerve through difficulty of movement of the lower jaw, or of the facial nerve through difficulty of lip movement, or in paralysis of the vocal cords through the recurrent laryngeal branches of the vagus nerve, or in lesion of the hypoglossal nerve by reason of paralysis of the tongue. Written or printed speech involves the optic nerve through vision, and deafness through the cochlear nerve may prevent speech being heard; various emotions as anger, fear or excitement may interfere with or prevent speech temporarily and phonation may be functionally lost in hysteria.

Bulbar Palsy.—Bulbar palsy, or glossolabiolaryngeal paralysis, is a disease involving the hypoglossal nerve probably to a greater extent than it does other nerves. It may form the terminal stage of amyotrophic lateral sclerosis or progressive muscular atrophy, or it may not be connected with these conditions.

Etiology.—It rarely occurs before mid-life and is usually a disease of old age. Some authors state that men, and others that women, are more frequently affected. In the author's experience men have been affected far more frequently than women. The cause of the degeneration is unknown. Syphilis seems to have no relation to the condition.

Symptomatology.—The patient usually begins by having some difficulty in pronouncing certain letters, especially d, j, k, l, r, s, t, and later speech may become so poor that it cannot be understood. The tongue is bilaterally paralyzed, cannot be protruded, is atrophied and shows reaction of degeneration. It lies flabbily in the mouth and fibrillary tremors may be detected. The lips are paralyzed, or partially so, and cannot be puckered and whistling becomes impossible. Saliva dribbles from the mouth and the lower face lacks in expression. Swallowing becomes more and more difficult. Liquids are more difficult to swallow than solids or semisolids and regurgitate into the nasopharynx. The muscles of the larynx are affected. The deep reflexes are usually increased and frequently the jaw jerk is pronounced. The patients are emotional and laugh or cry easily. Intelligence and sensation remain unimpaired. Toward the end the heart and respiration become affected.

Diagnosis.—In the diagnosis two conditions especially should be mentioned—pseudo-bulbar palsy and acute bulbar myelitis. *Pseudo-bulbar palsy* is a condition due to bilateral facial palsy, or to multiple cerebral lesions which may simulate bulbar palsy to a certain extent. In pseudo-bulbar palsy, however, there is no reaction of degeneration, no atrophy,

and fibrillary tremors do not occur. *Acute bulbar myelitis* is a form of polioencephalitis. It is an acute condition usually preceded by headache, malaise and fever in which there is difficulty in swallowing and paralysis of the tongue, palate and face with partial or complete paralysis of the vocal cords. Those affected with acute bulbar myelitis may make a prompt recovery but 50 per cent. die from the condition. *Hemiplegic lesions* may involve the tongue, but when this is the case there is no reaction of degeneration and no atrophy.

Treatment.—No treatment influences the course of the condition.

Prognosis.—The condition rarely lasts over three years and the patients die with cardiac and respiratory failure.

Pathology.—True bulbar palsy is due to a chronic degeneration of the motor cells of the pons and medulla without sensory cell involvement. The pyramidal tracts are always involved.

Spasms of the Tongue.—In epilepsy and chorea, and sometimes in hysteria, there occurs spasmodic movement of the muscles of the tongue. Spasms of the tongue may be tonic or clonic and occur in attacks. Many of these cramps or spasms are psychogenic in character and can be cured by psychotherapy. The author recalls a case of transient tonic spasm of the tongue in a syphilitic public speaker, which never occurred except during a speech and would last five or more minutes.

Aphasia.—Aphasia literally means the loss of the power of speech, but modern usage has divided the term into sensory and motor aphasia.

By *sensory aphasia* is meant an inability to understand written or printed words even without loss of the sense of vision and hearing. Sensory aphasia is considered in this article in the description of the optic nerve, under the headings of Visual Aphasia, etc.

Motor aphasia is an inability to speak because of some brain disturbance, although there is no paralysis of the muscles of articulation. Hemorrhage in the region of the middle cerebral artery is probably the most frequent cause of motor aphasia. The so-called Broca's convolution, part of the inferior frontal convolution formerly supposed to be the speech center, has been shown by Marie and others to be far too restricted, although the full extent and location of the speech center is still not definitely settled. In right-handed people the speech center is in the left cerebral hemisphere, or, at least, is functionally active on that side. Pure left-handedness is rare, and it is not established whether the speech center is ever entirely active in the right hemisphere in these individuals.

Aphasics usually are able to use one or two words, most frequently "yes" and "no," but by these words they have to express everything. Sometimes in association with musical memories whole verses can be sung when otherwise nothing can be expressed by speech. Patients with aphasia frequently know what they wish to call an object and know its usages, but are unable to call the name of the object shown them.

Most aphasics remain so, but with the reestablishment by collateral circulation of the areas cut off from sufficient blood supply in certain cases, or by reëducation, possibly of the opposite center, in young individuals the aphasia may disappear.

CLINICAL TESTS FOR DISTURBANCE OF THE CRANIAL NERVES

It is well now to consider certain brief clinical tests, in the routine examination of patients, which ascertain whether the individual cranial nerves are intact. If some disturbance is found, more elaborate and refined tests may be resorted to. The whole set of cranial nerves may be roughly tested in a few minutes, and the following suggestions are not intended to be more than a superficial means of eliminating cranial nerve disorder in the course of a general neurological examination.

Olfactory Nerve.—Ask the patient, "Is your smell very acute or very dull?" "Do you notice imaginary odors?" Blindfold the patient or make him close his eyes; occlude one nostril with your finger and put tobacco or coffee under the open nostril. Ask, "What do you smell?" Test the other nostril in same manner.

Optic Nerve.—Notice whether glasses are worn or not. To judge of the vision use a Snellen's test card at 20 feet distance, remove glasses if worn, cover one of the patient's eyes and ask him to read the smallest type he can see. Note amount of vision. Normally, number 20 line should be read at 20 feet. Try the other eye in the same manner.

Notice the pupils. Are they equal or unequal, dilated or contracted, regular or irregular in outline. Get the patient's face in a good light and cover both of his eyes with your hands, then remove your hands quickly and notice whether pupils react promptly or not, and then try each eye separately. Or take the patient into a dark room, or get his face in a shadow, and let the rays of a flashlight shine on each pupil separately in an oblique direction and notice whether or not the pupils contract.

Get the patient in a good light and with both of his eyes open ask him to look at a distance, then put your forefinger a few inches in front of his eyes and ask him to look at the end of your finger. Notice whether the pupils contract while thus converging.

Dilate the patient's eyes with a few drops atropin solution or 4-per cent. cocain solution. In fifteen minutes take him into a dark, or darkened, room, and look at his optic nerve head (disk) with the ophthalmoscope. Look for choked disks, pale disks, or disks with the margins blurred. Also look for hemorrhages, white spots, or other retinal changes.

In a good light make the patient close one eye, say the right, and fix the other, the left eye, upon your right eye, which should be about a yard distant. Move your right hand from the outer temporal region toward the nasal region, at the same time moving the fingers of this hand and keeping your hand equally as far from your own, and the patient's eye. Note, in comparison to your field, the temporal field of the patient which is indicated as soon as he sees your fingers moving. Try the same thing, with your right hand, from above downward and from below upward. With your left hand try from the nasal side toward a line from your eye to the patient's. These are tests for the

visual field and for hemianopia. With bits of colored paper you can test in the same manner for the color field. For refinement a perimeter should be used.

Oculomotor, Trochlear and Abducens Nerves.—Look for ptosis and note if the eyeball is looking in an outward and downward direction with the pupil dilated, which indicates oculomotor paralysis. Make the patient, with the eyes open, follow your finger moved in an upward, downward, inward and outward direction. If the trochlear nerve is paralyzed the motion upward will be limited and if the abducens is paralyzed the eye is turned toward the nasal canthus and will not follow the finger outward past the mid-line.

Trigeminal Nerve.—Inquire if there is pain on either side of the face and test sensation to touch on the face. A light stroke with the finger, or the corner of a handkerchief, will answer if cotton is not available.

Masseter Nerve.—Put your hands over each of the patient's masseter, or temporal, muscles and direct him to bite down and note if you feel them contract on each side.

Facial Nerve.—Note flatness of the muscles on either side of the face and observe whether the glossolabial folds are equal. Make the patient grin, close the eyes tight, wrinkle the forehead and note any loss of motion.

Intermediate Nerve.—The test for taste to the anterior two-thirds of the tongue, as indicated for the posterior part, will be found under Glossopharyngeal Nerve.

Cochlear Nerve.—Is the patient deaf? If so, is the deafness unilateral or bilateral? Is the deafness from a failure to hear sounds through air or bone conduction? With a watch or tuning fork note whether the ticking or vibration is heard well several inches from each ear while you occlude with your finger the ear not tested. In the same manner try the hearing with the watch pressed against the mastoid bone. With neither ear occluded press the watch, or tuning fork, against the center of the forehead and ascertain if hearing is as good in one ear as in the other. Does the patient complain of tinnitus?

Vestibular Nerve.—Has the patient nystagmus? Does the patient complain of vertigo? Make the patient turn around several times in one direction and then in the other and observe if this brings out excessive vertigo. Does the patient incline or lurch to one or the other side, or forward, or backward, in walking?

Glossopharyngeal Nerve.—Touch each side of the pharynx with a swab and see if sensation is preserved. Test taste to the posterior part of the tongue by making the patient protrude his tongue and touching one side and then the other with solutions of salt, sugar, quinin or vinegar in succession. Let the patient point, on a card marked "sweet—sour—bitter—salt," signifying which he tastes. Try one at a time and make him rinse his mouth with water before applying each solution. Also see that the tongue is fairly dry so that the solution will not run off the tongue and be appreciated by the taste bulbs of the mucous membranes of the mouth.

Pneumogastric Nerve.—Make the patient open his mouth and note whether the uvula is pulled to one or the other side. While his mouth is open make the patient say "Ah" and note if either side of the soft palate lags. Has the patient aphonia or hoarseness? With a laryngoscope notice whether the vocal cords contract normally. (Recurrent laryngeal branch.) Inquire into vagotonic symptoms—bradycardia, arrhythmia, spastic constipation, sweaty palms, nervus rigors and spells of apprehension.

Spinal Accessory Nerve.—Can the patient shrug each shoulder separately?

Hypoglossal Nerve.—Can the patient protrude his tongue? If so, is it protruded in a straight line? Is there atrophy of either side of the tongue?

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CHAPTER X

TRIGEMINAL NEURALGIA

(*Fothergill's Disease*)

BY CHARLES METCALFE BYRNES, B.S., M.D.

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Anatomy.—The trigeminal, or fifth cranial nerve, consists of a small motor root, a large sensory root, and the semilunar or gasserian ganglion, from the anterior border of which proceed the three main divisions of the nerve—the ophthalmic, the maxillary, and the mandibular branches. These three branches leave the cranial cavity through the sphenoidal fissure, the foramen rotundum, and the foramen ovale respectively, and make their superficial appearance upon the face at the supra-orbital notch, the infra-orbital foramen, and the mental foramen, as the supra-orbital, infra-orbital, and mental nerves. The first two branches contain only sensory fibers and innervate, in general, the skin of the brow, as far as the vertex, the eye, the cheek, the upper lip, the nose, the upper teeth and gums, and the nasal mucous membrane. The third, or mandibular division, is a mixed nerve, and contains all of the axones from the motor root, together with a large number of sensory fibers. The motor fibers innervate the muscles of mastication; while the sensory fibers convey impressions from a portion of the external ear, the anterior portion of the tongue, the mucous membrane of the cheek, the lower teeth and gums, and the integument of the lower lip and chin. The motor nucleus is situated within the tegmental portion of the pons, and the sensory fibers of the entire nerve have their cells of origin within the gasserian ganglion. For a more detailed account of the anatomy and distribution of the trigeminal nerve, the reader is referred to the Chapter on Diseases of the Cranial Nerves, and to the various textbooks upon Anatomy.

Definition.—"Trigeminal neuralgia" may designate a great variety of painful affections of the fifth nerve, or it may be used to indicate a particular affection of this nerve, in which pain is the essential feature. It is in this more restricted meaning that it is employed in the following pages to designate a clinical entity distinguishable from all other painful affections of the fifth nerve, and one in which the etiology

and pathology are undetermined. Not all fifth nerve pains are of the neuralgic type, nor are all neuralgic affections of this nerve, strictly speaking, trigeminal neuralgia. Migraine, the various cephalalgias, and the psychalgias are painful disturbances within the trigeminal area; and it is not uncommon for the layman, and sometimes even the physician to refer to these affections as trigeminal neuralgia. There are, also, many secondary affections of the nerve, dependent upon a demonstrable lesion within the cranium, at the base of the skull, or in the nose, sinuses, or teeth, which are similarly designated; and the pain induced by such lesions may even be confined to the anatomical distribution of the affected nerve, and thus, increases the difficulty of differentiating the two types of trigeminal pain. The attempt has been made to designate the two groups of neuralgic affections of the fifth nerve according to the intensity of the pain, as "minor" and "major" trigeminal neuralgia; but this distinction is not always well made, for the pain in both types may be excruciating.

Since an accurate definition of the disease cannot be given without describing the symptoms which serve to establish its entity, any effort to define the disorder would be merely a résumé of the following pages. It is to be understood, however, that throughout this chapter, the terms "trigeminal neuralgia" and "major neuralgia" unless otherwise stated, refer to the definite clinical disease only. All other neuralgias of the fifth nerve will be designated as minor neuralgias, or referred to in terms of their associated pathological condition.

Etiology.—The etiology, like the pathology, rests in obscurity, and space will not permit, nor would it be to any advantage to enumerate the various theories which have been advanced. The situation may best be summarized in the general statement that the essential diagnostic features of trigeminal neuralgia are its clinical manifestations and the inability, after a painstaking examination, to establish a demonstrable pathological or etiological foundation for the disease. If either one, or both of these fundamental aspects of the disease can be definitely determined in any individual case, the chances are that it is not one of genuine major neuralgia; but of necessity a secondary minor neuralgia, dependent upon the abnormality which has been discovered. It should not be inferred, however, that major trigeminal neuralgia has no etiology or pathology, but that present methods of examination have not furnished the means of establishing them.

Exposure to cold, sudden variations in temperature, the higher altitudes, emotional states, and overwork have been said to be responsible for the condition; but their causative relationship has not been established. Season appears to have no particular etiologic bearing, but when the disease is once established, the attacks are slightly more frequent during the winter months.

Among the infectious and constitutional diseases, syphilis, tuberculosis, gout, rheumatism, malaria, migraine, influenza, diphtheria, and erysipelas have been thought to be of some etiological importance. Except in rare instances, however, their relationship has been questionable.

Cachectic states, constitutional inferiority, a neuralgic diathesis, and even the cancerous acrimony of the older Fothergill have had their share of etiologic responsibility. Marked general anemia has been regarded as a cause by Rose; and Keen and Spiller have recorded an instance in which the disease developed in a female who had become anemic from uterine and rectal bleeding. Anstie has expressed a similar opinion in his statement that: "Neuralgia is the prayer of the nerve for blood," and the probable ischemic nature of the attack has been referred to in discussing the pathology of the disease. Brain tumor, and numerous extracranial and intracranial disorders are sometimes accompanied by trigeminal pain; but in these instances the clinical picture is usually atypical and serves to differentiate the disease from the genuine attack, although the local disorder may not, at the time, be demonstrable.

The rhinologist, laryngologist, and dental surgeon have diligently sought to establish the etiology in some local condition within their particular departments of activity. Thus, sinusitis, otitis, tonsillitis, pyorrhea, dental caries, alveolar abscess, and the pulp stones and osteodentine deposits described by Brownfield, Macmillan, and Goodwillie are given etiological prominence. Jobson has called attention to pressure upon the nasal septum by hypertrophy of the middle turbinate bone, and Moore has recorded an instance of trigeminal pain of twenty-five years' duration from inclusion of a foreign body in the right maxillary antrum following extraction of a tooth. A critical review of such cases will generally reveal a clinical picture quite distinct from that which is usually recognized as major neuralgia; and I know of no instance in which the genuine affection has been cured by any local operation upon the nose, throat, sinuses, or teeth. This should not, however, make one less careful to eliminate all possible sources of peripheral irritation in every case of trigeminal pain before arriving at a diagnosis of major neuralgia, or resorting to minor or major surgical interference.

Toxins of gastro-intestinal origin, and circulating toxins from obscure foci of infection, with a selective affinity for the trigeminal fibers, have been suggested as probable etiological factors, but their relation to the disease has not been established.

SEX.—The disease is slightly more frequent in the female, although the statistics of Head, Rose, J. Fothergill, and Patrick furnish no explanation for this preponderance. Patrick correctly concludes that the difference in the two sexes is probably negligible. Beckman is of the opinion that the two sexes are equally affected. In the writer's series of 108 cases there were 49 males and 59 females, and in 240 cases collected from the literature in which sex was mentioned, there were 118 in men, and 122 in women.

AGE.—It has been stated that trigeminal neuralgia is a disease of the "degenerative period of life"; that it begins most frequently during that period in which arterial and retrograde changes make their appearance. It is certainly more common during the fourth and fifth decade, although it is said to have begun as early as 17, and Patrick obtained a history of the onset at the ages of 7 and 8 in two of his

cases. The writer's youngest patient was 23 when the disease began. In the majority of instances, however, a history of an onset before the age of 30 should be regarded with suspicion.

RACE.—Apparently the disease presents no racial characteristics, but in the author's experience at the Johns Hopkins Hospital Dispensary, he has seen only two well-defined instances in the colored population. In fact, trigeminal pain in the colored man at once makes the diagnosis of major neuralgia questionable. In this race the symptoms are usually atypical and are generally dependent upon a syphilitic, nasal, or dental disorder.

HEREDITY.—Dana states that occasionally an hereditary history may be obtained. In one of Patrick's cases he obtained a history of the disease in the patient's mother, and in seven other instances in some antecedent or remote relative. He attaches, however, little significance to these observations, and Head denies altogether the importance of heredity. None of the writer's patients gave a history of the disease in the family.

INDIVIDUAL SUSCEPTIBILITY.—Much discussion has arisen concerning the relationship of the neuroses to the disorder. By some, it is maintained that those of a neurotic temperament are more susceptible; others deny the relationship. The author has not found the neuralgic patient particularly neurotic, nor has the history of an antecedent neurosis been generally obtained. It has also been suggested that erysipelas, remote or recent, is a predisposing factor, and in one of the author's cases the disease was said to have immediately followed an attack of erysipelas.

TRAUMA is said to have been a contributing factor in a few instances, but it probably has no prominent etiological relation.

Symptomatology.—Trigeminal neuralgia is not a rare disease, and is said to be only slightly less frequent than sciatica. Conrad's statistics from the Bonn clinic show, that among 717 cases of neuralgia of all types, 239 were of the fifth nerve, and Jelliffe found, among 613 cases of neuralgia in Starr's clinic, involvement of the trigeminal nerve in 315 instances. A history of antecedent disease is rarely obtained; the general health is usually good; the age of onset is rarely before thirty; and the patient gives a more or less characteristic history according to his intelligence and descriptive ability.

THE ONSET is usually without warning and is said to have occurred during a period of perfectly good health. While performing some trivial act, or while talking or eating, a sudden sharp pain is experienced in a particular area upon one side of the face, or in the gum, about the region of an upper or lower bicuspid tooth, but not necessarily in the tooth. In some cases the pain is described as darting, stabbing, or lightning-like from the beginning, and radiating throughout the length of the upper or lower jaw, but not necessarily associated with any particular facial movement. The interval before the next attack may vary from a few days to weeks or months; but, in time, recurrences are so frequent as to make the patient seek relief; and, since the teeth are not infrequently suspected, one or several of them are

generally extracted. In fact, the author has never seen a patient in which this measure was not adopted. Following this, there may be a period of relief of variable duration; but finally the disease manifests itself with its full intensity and characteristic features. In a few instances, a history is obtained of a more violent onset in which the pain is more intense and more widespread from the beginning. A local point of irritation may even be suspected in the teeth or gums, but it cannot be attributed to any one tooth or definite area of the gum; and there may be minor vasomotor or paresthetic disturbances of the affected part, immediately preceding the attack.

THE ATTACKS.—While the history of the onset is significant, the *character of the pain* and the unique features of the attack are diagnostic. The pain is of short duration, varying from $\frac{1}{4}$ to $\frac{1}{2}$ a minute, and rarely as long as two minutes, although Patrick states that he has observed a patient in whom it lasted fifteen minutes. Jelliffe is of the opinion that "A single attack may last a few days, or in the severer forms several weeks, the patient not being free from pain day or night." So long a duration is, however, extremely uncommon, and at once discredits a diagnosis of major neuralgia. It is therefore important, in securing the history, to inquire minutely into those cases in which the attack is said to be of more than two minutes' duration. It will usually be found that the patient has not timed the attack; that he has an exaggerated conception of its duration because of the intense suffering; that the attacks have recurred with such rapidity that one paroxysm has scarcely subsided before the next begins; and that he has regarded the series of attacks as one long continued pain. The author recalls an instance in which the attack was said to continue for an hour. He had the opportunity to observe the patient for that length of time, and found that during the period he suffered from forty-two distinct paroxysms, at intervals varying from fifteen to forty-five seconds. The short duration of the pain is further emphasized by the words with which the patient attempts to describe it. It is sometimes referred to as a shoot, stab, jab, knock, shock, dart, flash, or twinge; and to these Patrick has added the two very distinctive expressions, "zipp" and "bing." The same author has also directed attention to this feature of the disease in the statement that, "pain about the face, forehead, or temple which is a steady ache, even with exacerbations, or which is continuous for half an hour or more is not trifacial neuralgia." The pain is sometimes described as boring, burning, or throbbing, although the throbbing is not synchronous with the pulse.

The *short duration* of the pain, its *paroxysmal* nature, and *complete relief between attacks* are three distinguishing characteristics; and to these might be added, the *sudden cessation* of the attack. It disappears as suddenly as it commences. It is not only surprising but astounding to see a patient, whose expression exhibits every indication of his agony, suddenly breathe a sigh of relief and announce: "It's all gone!"

In *trigeminal neuralgia*, the pain must not only possess these characteristics, but it must follow the anatomical distribution of the affected

nerve. A correct interpretation of the course of the pain presupposes, therefore, knowledge of anatomy; and perhaps, in some instances, an incorrect diagnosis has been made because of an inaccurate knowledge of the anatomy of the fifth nerve, rather than from failure to recall this distinguishing feature of the attack. It is true that pain in any part of the face must necessarily be in the course of some branch or filament of the fifth nerve; but it must be more than this. It must be so definitely localized that there can be no question as to which branch is affected; and any pain in the face or head not thus definitely delineated is not trigeminal neuralgia. This aspect of the disease was well known even to the earlier writers upon the disorder, since the younger Fothergill(S.) has remarked that: "From an intelligent patient's description you might almost paint upon the external skin the ramification of the affected nerve."

The manner in which the patient indicates the location of the pain is characteristic, and is a most trustworthy index to the correct interpretation of symptoms. Were it permissible to arrive at a diagnosis from any one symptom of a disease, this feature of trigeminal neuralgia should unhesitatingly be awarded that distinction.

If a patient with any head pain, other than true trigeminal neuralgia, is asked to indicate the seat of pain, he will almost invariably use all of the fingers or the *entire hand*. If the pain is distributed over the entire half of the face, he will usually place the hand, without hesitancy, upon the forehead and carry it backward and slightly downward toward the ear, then forward into the region of the maxillary division, and again backward, downward and forward toward the chin, into the area of the maxillary nerve; or else the whole hand is placed upon the side of the head, in an effort to indicate the diffuseness and indefiniteness of the pain. In case the affection is confined to the region of either the second or third branches, the hand, *with all the fingers extended*, is placed upon the cheek, or over the lower lip and jaw, and held in this position, or else drawn backward toward the ear. In certain affections of the head, the pain is so widespread that it may not be definitely localized; and the patient may then make no effort to do so, or merely reply that "the whole face aches on one side."

The patient with trigeminal neuralgia has, however, a unique method of indicating the location of the pain, in spite of the teaching, that: "If the sufferer indicates the seat of pain by touching the face, a doubt that he has neuralgia at once arises." These patients will not infrequently indicate the seat of the affection, but in an entirely different, and more cautious manner from that observed in those who suffer from the other types of cephalic pain. If, as has been suggested, there is fear of touching the face, the hand, *with the index finger alone or with the index and middle fingers extended*, may be brought slowly and carefully toward the face and pointed at the seat of pain. In many instances, however, the patient will have courage enough to touch the face, but with only one or two fingers, instead of the whole hand, and will then definitely outline the course of the affected nerve. The author

has never seen one suffering from major neuralgia of the fifth nerve indicate the location of the pain by using the whole hand, or exhibit any indecision as to the location, intensity, and demarcation of the pain. This peculiar manner of indicating the seat of pain is due to the fact, that in the great majority of cases, the paroxysm is induced by various forms of peripheral irritation; and usually, as Anstie has remarked, by an exciting cause which is trivial in comparison to the severity of symptoms. Thus a light touch, as from a wisp of cotton, a lock of hair, or the draft created by closing a door are, as a rule, more apt to initiate an attack than are the firmer degrees of pressure. In fact, firm pressure is said to sometimes diminish the pain. Eating, talking, hot or cold drinks, a sudden jar, a misstep, riding, driving, using the handkerchief, smoking, shaving, touching the cheek, lips, or gums with the tongue, and contact of the bedclothes during the night are, in many instances, sufficiently irritating to induce an attack. A patient, who was particularly susceptible to currents of air, but who enjoyed his afternoon drive, came to the writer's office with his entire head, except the eyes and nose, enclosed in an especially constructed woolen mask. Certain movements of the facial muscles, jaw, or tongue are at times especially irritating, so that the patient soon learns to avoid them, and is forced to make his wants known by signs or in writing.

Irritation of certain areas upon the face, especially at the angle of the mouth or ala of the nose will, in many cases, almost invariably precipitate an attack. Often, however, it requires a particular kind of movement or tactile impression in these regions to induce the paroxysm, and the author recalls such an instance in which the pain was confined to the maxillary nerve. At times, the attack occurred spontaneously and the upper lip, nose, and cheek could be rubbed, stroked, or kneaded with impunity; and light touch was easily tolerated so long as the direction of the stroke was downward from the eye toward the lip. If, however, the lightest stroking were directed from the lip upward, or from the ala of the nose toward the outer canthus of the eye an attack was almost certain. Patrick has happily designated these contact areas as "trigger zones," or "dolorgenetic zones," and has called attention to the fact that they may, at times, be situated upon an area of the skin surface which is innervated by one of the trigeminal branches not implicated in the attack. This experience was confirmed by several patients in whom an attack, confined to the maxillary nerve, was induced by touching the lower lip or chin, or in whom a supra-orbital pain was started by irritation of the perfectly normal maxillary area. In rare instances the dolorgenetic zone is said to be situated in some portion of the body, other than the head or face; and while it is not uncommon, during a severe paroxysm, to have the pain radiate to the neck or occasionally down the arm, Patrick has observed two instances in which the attack was induced by using the arm upon the affected side. He offers no explanation for this unusual observation, nor does he speak of the arm as a possible dolorgenetic zone. Perhaps the attack in these two instances might have been due to a very slight

associated movement of the facial muscles, or to a certain degree of tension upon the adjoining skin surfaces of the shoulder, neck, and face. No doubt, these trigger zones were, for a time, misinterpreted, as Valleix points, which are said to be present in trigeminal neuralgia; but the author's experience is in accord with the opinion expressed by Patrick, that Valleix points, as such, are not present in the typical disease. Rose thinks they may be present, but are not common. S. Fothergill claims that the attack is also induced by the emotional states of passion, anger, and grief; and the author has upon several occasions observed an apparent relationship between the two.

That the paroxysms are, sometimes, accompanied by *movements of the facial muscles* cannot be questioned; but the significance of these contractions, and their position in the clinical syndrome have been the subject of dispute. By some, the spasm is regarded as involuntary and forming an essential part of the disease, which led to the designation of the affection, by the French, as *tic douloureux*, or painful spasm. Others consider the spasm as voluntary, and merely a secondary feature adopted as a means of relief. During a severe paroxysm, the movements are not confined to the face; and the patient will sometimes walk, clench the hands, grasp an object, beat the head, or even roll on the floor; or he may chew continuously, suck the cheeks, or make blowing movements, and at the same time rub the face violently. One of the writer's patients always carried a pad, made of rough cloth, which could be slipped over the right hand like a mitten. Upon one occasion, while conversing in perfect comfort, he suddenly remarked: "It's coming," and withdrew the pad from his pocket just in time to begin rubbing the face violently before the attack reached its greatest intensity. He continued this performance throughout the duration of the paroxysm, and quietly put his pad away for the next occasion. The use of this measure, during many months, had so irritated the skin of the entire half of the face that it was firm, glossy, and hyperplastic. The frequent occurrence of these widespread motor disturbances has contributed to the belief that the entire motor features of the disease are voluntary. The author has never felt convinced that the simpler facial movements are, strictly speaking, a part of the disease and involuntary; and similar opinions have been expressed by S. Fothergill, Patrick, and Beckman.

Objective sensory changes, other than occasional hypersensitiveness, are not a part of the disease; and an actual loss of either tactile pressure or thermic sensibility is never present. *Pain in the distribution of any or all of the branches of the trigeminal nerve associated with objective loss of cutaneous sensibility in any of its elements is not major trigeminal neuralgia.*

During the height of a severe paroxysm, certain vasomotor and secretory disturbances have been observed. The face is said to be congested, swollen, and edematous; and in some instances, immediately preceding the attack, there may be a hyperemic streak upon the skin which indicates the course of the affected nerve. Excessive lacrima-

tion, suffusion of the conjunctiva, nasal discharge, salivation, and local sweating have also been described. Keen and Spiller record an instance in which the attack was accompanied by excessive lacrimation, and the discharge of a bloody, yellowish fluid from the mouth. The author has the history of a patient who, following a recurrence after the major ganglion operation, claimed that the attacks were associated with a discolored, foul-smelling oral discharge. She was referred to a competent laryngologist for consultation, but neither he nor the writer was able to verify her statement, nor did she ever comply with the request to bring a specimen of the sputum.

Major trigeminal neuralgia, then, with the exception of the associated motor, secretory, and vasomotor phenomena, is distinctly a subjective disorder in which a particular kind of pain is the distinguishing feature. The physical examination of the patient, the investigation of the reflexes, and the laboratory findings are entirely normal, or else reveal the changes which are to be expected in those of advancing years. Moderate thickening of the peripheral and retinal arteries, hypertension, and a slight degree of albuminuria may be present in the older patients; but, with the exception of arteriosclerosis, the author has never felt that even the most advanced senile changes had any particular relation to the disease. The Wassermann test has been consistently negative, and the x-ray examination has failed to reveal any abnormality.

SPECIAL FEATURES OF THE DISEASE.—The disease is essentially unilateral, although both sides of the face are occasionally involved. It is said to be more common upon the right side; that the second or maxillary branch is most often affected; and that the attacks are more frequent during the day than during the night. Earlier students of the disease, the two Fothergills, Campbell, and Nasmyth, as well as those of a later date, have attached some importance to the nocturnal and diurnal features of the attack, in an effort to differentiate the disorder from other face pains which were supposed to be of rheumatic origin. While a decision in the matter may be of little importance, opinion of to-day is distinctly in favor of the diurnal frequency of the paroxysms. This preponderance of daily paroxysms is thought to depend upon the more numerous forms of external irritation to which the patient is subjected in his waking hours. Attacks during the night are usually attributed to contact with the bed-clothes, or changes in position; and there are a few instances in which the patient is awakened by a severe spontaneous paroxysm, although as a general rule sleep is undisturbed. Both sides are probably more frequently affected than one is led to believe from isolated reports. As early as 1804, S. Fothergill, recognized the bilateral affection, and refers to two instances recorded by Foquet and one by Pujol. Patrick in a series of 220 cases found the affection bilateral 15 times—a rather high percentage—but, as he remarks, some of these were at first unilateral and only later became bilateral. In only one instance did he observe both sides affected at the same time. Vaughn records an instance in which both sides were involved, but not simultaneously at the outset. The author has seen the

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condition bilateral 4 times and has collected 19 instances from the literature; but in none of these were the two sides involved at the beginning, or at the same time.

Of the two sides of the face, the right is said to be more frequently involved, although no satisfactory explanation has been given for this predilection. Statistics upon this feature of the disease are unreliable unless based upon a large number of cases and extensive experience. During a certain period of Patrick's experience, and likewise during the writer's, there was a preponderance of the right-sided involvement. Thus, in the latter's first ten cases, seven were affected upon the right side; and in Rose's early experience, the right trigeminal nerve was involved in every case. From the statistics of Tinker, Patrick, Foquet, Pujol, and others, the writer has collected 335 cases in which the affected side and bilateral involvement are recorded. Of these, the right side was involved in 205 instances, the left in 111, and both sides in 19 cases. In the writer's series of 108 cases, there were 63 upon the right side, 41 upon the left, and 4 in which both nerves were involved. Thus, in a total of 443 cases, the disease was confined to the right nerve in 268 cases, to the left nerve in 152 cases, and affected both nerves in 23 cases.

Neuralgia of the Ophthalmic Nerve.—Supra-orbital neuralgia is confined largely to the frontal division of the nerve, and is familiarly known as brow ache, from its supposedly malarial origin, and the occasional periodicity of the attacks. The ophthalmic nerve is not infrequently involved in diseases of the frontal sinus or orbit, in herpes, and in syphilis of the nervous system; but as an isolated affection in major neuralgia it is extremely rare. Dana, in a series of 45 cases of trigeminal neuralgia, found the affection confined to the first branch in only one instance. In the author's series, the nerve was involved alone 12 times; but none of them was characteristic of the major attack. In two instances the affection was secondary to gangrenous herpes; in four, there were migrainoid features; in three the attacks were thought to be psychic; and the remaining three cases possessed features essentially different from those seen in genuine neuralgia. Patrick is of the opinion that, of the three branches of the trigeminal nerve, the first "is by far the least frequently affected"; and that "pain limited to the supra-orbital region probably is not neuralgia at all." The nerve is sometimes affected in conjunction with neuralgia of the second or maxillary division, but no instance has been recorded of involvement of the first and third branches alone. Occasionally, the supra-orbital attack may be quite like that observed in genuine neuralgia, and there may be a dolorigenetic zone at the side of the nose or on the upper lip. It has been suggested that such cases are not primary affections of the ophthalmic nerve, but that the disease really began in the maxillary area, subsided, and after a variable interval, during which the primary affection may have been forgotten, recurred in the supra-orbital region.

Neuralgia of the Maxillary Nerve.—Statistics vary as to the frequency with which the disease is confined to the second division of the trigeminal nerve. Dana, J. Fothergill, S. Fothergill, and Camp-

bell are of the opinion that the maxillary nerve is more frequently affected than the mandibular nerve. Head, Peet, and others have found the mandibular branch more often involved in the beginning, and a combined affection of the second and third branches to be more frequent than the involvement of either branch alone. Patrick, however, observed only slight predominance of maxillary involvement, and almost an equal percentage of isolated and combined affections of the maxillary and mandibular branches. In the author's series, the disease was confined to the maxillary nerve in 36 instances, to the mandibular nerve in 31 instances, and the two branches were simultaneously involved in 21 instances. In 40 additional cases, collected from the literature, the two branches were about equally involved, and both nerves were affected at the same time in 10 cases.

Neuralgia of the Mandibular Nerve.—This, the largest of the three divisions of the fifth nerve, consists of three important branches: the inferior dental, the lingual, and buccal nerves. Any one or all three branches may be the seat of neuralgia. The inferior dental nerve is affected about as frequently as the maxillary nerve; but pain in the lingual branch is comparatively rare. In the author's series of cases it was involved 5 times, and almost always in conjunction with the dental nerve. He has seen only one instance in which the disease was confined to the lingual nerve, and in this instance there was a definite trigger zone about the mental foramen. Bunting did not find it affected in any of his cases.

It has been stated recently, that among the unpleasant consequences following division of the trigeminal motor root there is paralysis of the buccinator muscle. This muscle receives its innervation from the facial, or seventh cranial nerve, and although filaments of the buccal nerve ramify through the substance of the muscle, no motor fibers are contributed to the muscle by this nerve. The buccal nerve is purely sensory, and innervates the mucous membrane of the cheek; and its course may be roughly outlined upon the face, by a line drawn from the tragus to the angle of the mouth, midway between the maxillary and inferior dental nerves. It is reasonable to suppose, then, that the buccal nerve might be subject to neuralgic affections, and upon one occasion the writer observed a patient who suffered from attacks limited to this nerve. The history and paroxysms were characteristic. She complained of pain in the cheek, and indicated an area upon the face corresponding to the course of the nerve. An attack was almost invariably induced by touching the inside of the cheek with the tongue, and the pain seemed to be *in* the cheek rather than upon the surface. There had never been pain about the upper lip, nose, lower jaw, lip, or tongue; nor was a paroxysm induced by irritating any of these areas. Since the pain seemed to be slightly nearer the upper than the lower part of the cheek, the attacks were thought to be due to involvement of the maxillary nerve, and it was accordingly injected with alcohol. Complete anesthesia was obtained in its entire distribution, but unfortunately the pain persisted, and the writer concluded that the affection might be

located in the buccal nerve. An injection was then made at the foramen ovale. This was followed by complete relief and anesthesia of the mucous membrane of the cheek, but the lower lip was only slightly numb and the tongue retained its normal sensibility.

Neuralgia of All Three Branches.—Simultaneous involvement of the three trigeminal branches is, with the exception of supra-orbital neuralgia and the bilateral affection, the least frequent clinical manifestation of the disease. It is even more exceptional to find the entire trigeminal area affected in the beginning, and Sicard states that: "All neuralgia which affects at its very beginning all three branches is not essential neuralgia." In the 335 cases of trigeminal neuralgia collected from the literature, there are only 28 in which all three branches were affected; and to these may be added 4 from the author's personal records.

Diagnosis.—Trigeminal neuralgia, in its major form, is a distinct clinical entity. The diagnosis rests entirely upon the clinical symptoms of the disease, and the inability to demonstrate any general or local condition upon which they depend. There should be no difficulty in recognizing the disease if the following essential features are observed: (1) The disease rarely begins before the age of thirty. (2) The patient usually has the appearance of general good health. (3) The pain must be of short duration; it must follow the anatomical distribution of the affected nerve; and it is of sudden onset, paroxysmal, and sharp, shooting in character. (4) Complete freedom from pain during the interval between the attacks, and the sudden cessation of the attack. (5) The attack is usually started by some form of peripheral irritation, though it may occur spontaneously. (6) The manner in which the patient indicates the seat of pain, or his unwillingness to do so. (7) There is no loss of sensation in any part of the trigeminal area.

Not infrequently, obscure painful affections of the fifth nerve are, from carelessness or inexperience, incorrectly diagnosticated as major neuralgia. It is, therefore, equally important that the physician should train himself to recognize what is not trigeminal neuralgia; and if any one of the following negative statements is regarded, it may be of service in the process of exclusion: (1) Any pain in the face which begins as a dull ache, increasing in intensity and gradually subsiding, although it may occur in paroxysms, is not trigeminal neuralgia. (2) Any pain in the face of more than two or three minutes' duration is, as a rule, not trigeminal neuralgia. (3) If there is any doubt in the patient's mind as to the location of the pain, or if the pain is so diffuse that its location cannot be definitely indicated, it is not major neuralgia. (4) Assuming the physician's knowledge of anatomy to be accurate, should the location of the pain be so indefinite that a decision cannot be reached as to which branch is involved, it probably is not trigeminal neuralgia. (5) If during a paroxysm, the patient can assume an indifferent attitude toward the situation, or if the attack can be aborted by any mental process, it is not trigeminal neuralgia. (6) Pain in the face associated with involvement of other cranial nerves is not major neuralgia. (7) Trigeminal pain accompanied by diminution or loss of

cutaneous sensation in any part of the fifth nerve is not genuine neuralgia.

DIFFERENTIAL DIAGNOSIS.—In the typical disease there should be no difficulty in making a diagnosis, for the major attack resembles nothing else. Perhaps the more frequent error, therefore, is made in diagnosing all varieties of face pains as major neuralgia, rather than in diagnosing the genuine attack as some other disease; and, no doubt, the greatest difficulty is experienced in the differentiation of the several types of trigeminal pain to be described in the following paragraph as:

Minor Neuralgias of the Fifth Nerve.—To this class belong most of the face pains accompanying diseases of the teeth, orbit, sinuses, ear, nose, and antra, and the painful symptoms associated with intra- and extracranial new growths, Meckel's ganglion syndrome, syphilis of the fifth nerve, and herpes facialis. Trigeminal pains are sometimes complained of in tabes dorsalis, multiple sclerosis, disease of the optic thalamus and other central lesions; but these pains are generally so unlike those experienced in neuralgia and so readily distinguished by their associated symptoms, that they may not properly be referred to as minor neuralgias. Space will not permit a full discussion of the differentiating features of each of these conditions, and though they may exhibit features which resemble the major attack, there is generally some atypical or additional symptom which enables one to conclude that the attack is not genuine. Not infrequently, there is diminution or loss of sensation in the area of the affected nerve, and Dana is of the opinion that progressive loss of sensation in the trigeminal region is usually unaccompanied by severe pain. This, however, is not uniformly true, for the writer has recorded an instance of progressive syphilitic paralysis of the fifth nerve in which loss of sensation was associated with excruciating pain. In many instances a local disorder is demonstrable, but occasionally the pathological condition may be obscure. It is then wiser to regard the condition as a minor neuralgia, and await further developments before resorting to minor or major operations upon the trigeminal nerve.

Hemicrania in which the gastric and ocular symptoms are wanting may sometimes be confusing, and is occasionally mistaken for supra-orbital neuralgia; but in hemicrania, the duration, character, and distribution of the pain, the history of a similar disorder in the family, and the periodicity of the attacks should enable one to differentiate the two conditions.

The *psychalgias of hysteria* and the *minor neuroses* are sometimes mistaken for major trigeminal neuralgia, and the writer knows of several instances in which these affections have been treated by minor or major surgical operations upon the trigeminal nerve without securing the slightest relief. These neurotic individuals generally give a history of a preceding nervous collapse or general ill-health, associated with or followed by pain in some part of the trigeminal area. Pain finally becomes the dominating symptom, and the patient seeks relief in ill-advised

operations upon the teeth, nose, or pharynx. The painfulness of the operation is added to the preëxisting pain memory, and finally a diagnosis of trigeminal neuralgia is made. There is another type in which the operation furnishes the mental nucleus of a subsequent psychalgia in those of the neurotic temperament. A minor surgical procedure or repeated painful treatments are resorted to for the relief of some simple affection of the head. Treatment is discontinued, but a vague indefinite pain persists. The neurosis asserts itself; apprehension and doubt arise; permanent damage is suspected; and a full-grown psychalgia develops. Hate, fear, and malice are liberally distributed upon the physician and all the circumstances associated with the operation. A careful inquiry, then, into the origin of the psychalgia, the long duration of the attack, and its subjugation to appropriate psychic measures serve to distinguish it from major neuralgia.

Complications and Sequelæ.—Trigeminal neuralgia does not predispose to any other disease, but the difficulty experienced in eating may lead to gastro-intestinal disturbances, malnutrition, and, in some cases, to extreme emaciation. Usually, the intervals between attacks are of sufficient duration to permit the reëstablishment of such functions as may have suffered during a prolonged series of paroxysms. It is claimed that pain is an important factor in the development of arteriosclerosis, and many neuralgic patients exhibit variable degrees of arterial thickening or hypertension. Whether the arterial changes are primary or secondary is still a matter of dispute, and those who regard them as primary, argue that trigeminal neuralgia rarely begins before the degenerative period of life.

Long suffering is rarely endured without a degree of mental and moral disorganization, and the neuralgic patient has sometimes sought relief in self-destruction, although the percentage of suicides is comparatively small. Peet, however, claims that it is greater than the mortality from the major operation. The author has not known the disease to terminate in this manner, and it is surprising how few patients exhibit even a moderate degree of depression. S. Fothergill has described melancholia and mania as rare complications, and morphinomania has been recorded in a few instances.

The vasomotor and secretory symptoms accompanying the attack are sometimes said to persist during the interval between paroxysms, and may therefore be properly referred to as complications. Among these, Rose has observed permanent inflammatory hyperplasia of the lips, cheek, tongue and floor of the mouth; and, occasionally, the skin upon the affected side is said to become thin, shiny, glossy, and atrophic from violent friction and the use of local applications. The tongue, upon the affected side, is sometimes coated or even furred, and Anstie concluded that the changes were trophic, or perhaps due to the fact that the affected side of the mouth is not used in chewing. Conjunctivitis, corneal ulcers, frontal periostitis, unilateral lacrimation and sweating, nasal flux, impaired hearing, defective taste, transitory blanching of the hair, paralysis of the third nerve, and erysipelas have been

described as complications of the disorder. Trophic retinal changes, facial paralysis, and spasms of the muscles of the jaw and neck have also been added to the clinical picture; and Patrick states that in one of his cases the teeth dropped out after the onset of the neuralgia.

Association with Other Diseases.—Major neuralgia is rarely associated with other diseases, but Dana and Putnam have attached special importance to its occurrence in conjunction with migraine. The two diseases sometimes occur together, and not infrequently a history of migraine may be obtained in the patient or in a relative, but the relation between the two is doubtful. Malaria has been thought to be a frequently associated disease, but the writer never saw a case of genuine neuralgia in which the history or symptoms indicated the presence of malaria, or in which the plasmodium could be demonstrated. Anstie, in his Lettsomian lectures, states that neuralgic patients are particularly susceptible to erysipelas, and that he has observed this disease in association with trigeminal neuralgia. One of the writer's patients also gave a history of the development of typical major neuralgia immediately following recovery from erysipelas, but the relation between the two diseases is probably coincidental. Robinson records a case of trigeminal neuralgia associated with Ménière's disease, and the author has seen a patient, with neuralgia on the right side of the face, in whom the attacks were frequently either preceded, associated with, or followed by the typical symptoms of Raynaud's disease in the right arm. Trigeminal neuralgia and the neuroses are sometimes said to be associated, but the neuralgic patient is only slightly more neurotic than the average individual, and the relation between the two diseases is probably negligible. Pyorrhea and syphilis may be coincidental infections, and a history of epilepsy, the psychoses, or mental deterioration is said to have been obtained in a few instances.

Treatment.—**DRUGS.**—Many unsuccessful efforts have been made to treat the disease by the use of *drugs*, and a great many medicinal substances are recommended in the earlier literature of the disease. **Quinin, arsenic, gelsemium, aconite, antipyrin, phenacetin, hemlock, cannabis indica, nitroglycerin, the bromids, the salicylates, strychnin, and castor oil** are some of the more widely used preparations, and Tracy reports some success from the use of **pilocarpin**. Most of the so-called **alteratives** and **antispasmodics**, the **sedatives**, and the **analgesics** have been administered with variable degrees of disappointment. **Aconite** has, however, enjoyed a fair reputation among a small number of reputable physicians, and **quinin** is thought to possess particular virtue. **Opium** is mentioned only to be condemned.

Those who have thought the disease to depend upon a form of intestinal intoxication have acquired a fondness for the use of castor oil in repeated and, at times, in heroic doses; and for a while the "**castor-oil treatment**" was favorably regarded. Hamill and Patrick have reported some success with the method, and Moore recommends its use in a modified Towns-Lambert course of therapy. He begins with a dose of **blue mass, or compound cathartic pill, and an enema.**

Atropin is then given every two hours until the physiological effect is obtained. Following the sixth dose of atropin, four to eight ounces of castor oil are administered, and repeated in from eight to twelve hours. After thirty or sixty hours of this combined atropin and castor-oil dosage, mucus is said to appear in the stools, and the atropin is then discontinued; but the oil is given twice daily for two days, and thereafter every other day for a week. During the treatment, the patient is kept comfortable by the use of **analgesics**, although it is claimed that the paroxysms begin to diminish in frequency and intensity after the third day.

Until within the last decade, **strychnin** was widely used with unquestioned success in a few instances, and with a certain degree of relief in a fair proportion of cases; and there are to-day some physicians who, in spite of the more modern methods of treatment, prefer to give the patient the benefit of a thorough course of strychnin therapy before resorting to other measures. In 1896, Dana reported that a certain number of cases of trigeminal neuralgia were curable by a method of treatment which he then introduced, with acknowledgment of his indebtedness to Dr. G. R. Elliot for suggesting the use of strychnin. The method consists of three essential procedures: (1) the hypodermic injection of massive doses of **strychnin**; (2) the administration of **potassium iodid** and **tincture of iron**; and (3) **rest in bed, light diet, and diuretics**. He cautions that the treatment admits of no halfway measures, and to be effective it must be given in full. The strychnin is given in single daily doses, beginning with $1/30$ grain (0.002 gram), and increasing slowly, until by the fifteenth or twentieth day from $1/6$ to $1/4$ grain (0.0108 to 0.016 gram) is administered, although few patients can tolerate more than $1/5$ grain (0.013 gram). This maximum dose should be continued for a week or ten days, and gradually reduced, so that by the end of the fifth or sixth week the initial dose is resumed. **Potassium iodid**, grains v (0.324 gram), together with **tincture of iron**, minims v (0.30 c.c.), are given three times daily, and both are increased, until grains xx (1.3 grams) of the iodid and minims xxx (2.0 c.c.) of the iron are being taken. **Nitroglycerin** is sometimes added to the potassium iodid. Rest in bed for four weeks is insisted upon, and two weeks more are usually required for a gradual return to mental and bodily activity. Should the pain return, the treatment is promptly resumed.

It is claimed that the larger doses of **strychnin** have an anodyne effect in which the patient is quieted for hours, but that sometimes the pain is temporarily aggravated. Robinson confirms the latter observation and accordingly condemns the use of strychnin. Dereum, however, in discussing this subject before the Philadelphia Neurological Society, 1914, spoke favorably of the use of strychnin, and it has been used by others with a measure of success. Since many of the writer's patients had already been subjected to a course of strychnin, and because of preference for other methods of treatment, the writer had no experience with Dana's method. It seems, however, that in early cases, or

as a temporary measure until other procedures can be employed, it may be adopted with some degree of confidence.

ELECTRICITY.—Attempts have been made to introduce a solution of the medicinal substance into the offending nerve by **ionization** or **electrolysis** according to the method advocated by Le Duc; and Turner records the successful use of this method in six cases. **Quinin** or the **salicylates** are the drugs usually employed; and the positive electrode, wrapped in absorbent cotton, is dipped into a solution of the desired drug and applied over the exit of the nerve. The current is then gradually increased to as much as ten milliamperes, and allowed to flow from ten to twenty minutes, when it should be gradually withdrawn. The writer has used the method in one instance of supra-orbital pain with gratifying results, but has had no experience with it in major neuralgia.

Electricity, in the form of the **x-ray**, **high frequency currents**, the **galvanic current**, and the **violet-ray**, has also been employed, and it is said to be of some value as a means of temporary relief. Wolf claims to have had a measure of success, in certain types of neuralgia, from thermopenetration or diathermia with the high frequency currents. Attempts have also been made to destroy the nerve by the **electric cautery**, according to the method of Tansini; and in the record of seventeen cases treated in this manner a period of relief, ranging from two to nine years, is said to have been obtained. Six additional cases have been treated by d'Este, who remarks that the ganglion may also be successfully cauterized. The method consists of pulling the nerve out of the foramen, introducing the electric needle into the canal, and allowing the current to flow from 5 to 10 seconds.

LOCAL APPLICATIONS.—Although it has been stated that "a patient who has been rubbing the face with liniments has something besides the major neuralgia," the earlier literature of the disease contains many references to the use of *local applications*. S. Fothergill recommends a novel and ludicrous method of local treatment when he advises that "the steaming entrails of a young pigeon be thrown into the face of the sufferer." (No doubt an effective measure in cases of hysterical psychalgia!) Of the more modern methods, **blisters**, **liniments**, **hot air**, **ethyl chlorid spray**, and **whisky**, held within the mouth, have been tried with uniform disappointment; and one patient-informed me that whisky invariably induced an attack. **Massage** and **hydrotherapy** in the form of sulphur baths or as Nasmyth suggested, dashes of **cold water** against the face until the teeth chatter, have been resorted to unsuccessfully. Climatic conditions, especially low altitudes and inland moderate temperatures, have received but little recognition.

TREATMENT OF MENTAL CONDITION.—The mental condition of the patient may be either harmful or beneficial. From the earlier writings up to the present time it has been conceded generally that worry, shock, anxiety, grief, and deep emotions have a harmful effect upon the disease, and not infrequently induce the paroxysm when once the disease is established. On the other hand, neuralgia is said to have been relieved by **mental diversion**. Rose states that cases are on record in

which a sudden cure has followed the reception of some unexpected mental or physical shock; and S. Fothergill records an instance in which a cure followed an interesting game of cards. It seems, however, that in such cases the diagnosis of major neuralgia is questionable. **Hypnotism** and **animal magnetism** were also advocated by earlier writers.

INJECTIONS INTO NERVE TRUNK.—With the exception, then, of **strychnin**, **castor oil**, and **electricity**, the medical treatment of the disease is largely of historical interest only, and the necessity for **surgical interference** or the adoption of measures directed toward the interruption of the nerve impulse is recognized by all who have had any experience with genuine major neuralgia. The majority of the author's patients had previously been treated by the various medical procedures with uniformly unsatisfactory results; and among his records are the histories of those who have tried **strychnin**, **castor oil**, **gastric lavage**, **electricity**, **climate**, **spas**, **hydrotherapy**, **osteopathy**, the various **tonics**, **sedatives**, **morphin**, and **Christian Science**.

This general inefficiency of most drugs soon led to the early adoption of various minor surgical measures, and finally to the more extensive operations of **Rose**, **Hartley**, **Krause**, **Keen**, **Cushing**, and others. The temporary relief obtained from the minor nerve operations, and the earlier high mortality following the ganglion operation have been constant stimuli in the search for a simple but effective method of treatment.

Within the past fifteen years, the *injection of physical or chemical substances into the nerve trunk*, and more recently into the **gasserian ganglion**, has occupied an important position in the treatment of the disease. At first the injections were made into the exposed nerve or ganglion, but a technic has since been perfected by which the injection may be made without exposing the ganglion or its branches.

Two classes of substances have been injected. First, those by which it was hoped to secure dissociation of the nerve fibers without necessarily destroying them; and for this purpose **air**, **serum chlorure**, **serum glucose**, **fibrolysin**, **cocain**, **methylene**, **strychnin**, **adrenalin**, and **magnesium sulphate** have been used. **Sicard** has found that the duration of relief following these injections varies from a few days to a few weeks. Secondly, destruction of the nerve fibers and subsequent **Wallerian degeneration** have been accomplished by the injection of substances usually employed to "fix" the nerve. Of these the more important are **osmic acid**, **the chromates**, **glycerin**, **formal**, the **salts of quinin**, **carbolic acid**, **chloroform**, **ether**, and **alcohol**; and while all of these produced the desired effect, many of them were followed by local necrosis and were subsequently abandoned. **Alcohol**, however, seems to have stood the test of time, and has become generally adopted as a substitute for all forms of peripheral operations.

Since the introduction in 1903 of **Schlosser's technic of injecting alcohol**, this method, with its several modifications, has supplanted all others. Its value has been attested to by the experiences of **Levy** and **Baudouin**, **Sicard**, **Harris**, **Kiliani**, **Ostwald**, **Härtel**, **Patrick**, **Grinker**,

and many others in this country and abroad. Space will not permit a full review of the development of the method, or the exposition of a technic in which special training and skill are essential. In an earlier paper upon this subject, the author has given an account of the anatomical and clinical results following the injection of alcohol; and a good account of Schlösser's technic will be found in the writings of Levy and Baudouin, Patrick, and Harris. It is important that something more than a mere literary knowledge be obtained; and it is strongly advised that the necessary skill and confidence be acquired in the anatomical laboratory before an attempt is made to treat the living subject. For these reasons, only such knowledge as may be necessary to enable the practitioner to form an intelligent conception of the procedure will be discussed in this chapter.

Schlösser, an ophthalmologist, found that the subcutaneous injection of 70 to 90 per cent. alcohol into the supra-orbital nerve immediately relieved the neuralgic attacks confined to this nerve, and similar injections at the infra-orbital and mental foramina produced equally good results. He subsequently developed a technic whereby the injection could be made with a long graduated needle, and without a cutting operation, into the trunks of the individual branches of the fifth nerve as they make their exit at the base of the skull. By this means the chemical destruction of the nerve was accomplished at a point much nearer its ganglionic origin, and, therefore, more lasting relief was to be expected. It was soon found, however, that the deep injection of the ophthalmic nerve, because of its proximity to the orbital contents, was entirely too hazardous even when practiced by the most skillful; and it has since been abandoned. In many instances Schlösser thought it advisable to give both the superficial and deep injection to make sure that the offending nerve had been successfully treated. Several modifications of his original technic have since been devised, and the injecting fluid has also been slightly altered according to the preference of the individual operator. Perhaps the most widely used method is that described by Levy and Baudouin, and while **simple ethyl alcohol** of the percentage mentioned is quite effective, **chloroform**, **ether**, **adrenalin**, and other substances have been added to the mixture.

The Injection.—With the patient lying down, and preferably in a hospital, the area selected for introducing the needle is painted with iodine, and rendered anesthetic with the ethyl chlorid spray, or by infiltration with Schleich's solution, novocain, or stovain. All the necessary aseptic precautions are to be observed throughout.

The treatment is necessarily painful, but probably not more so than a severe paroxysm of neuralgia. Some patients complain of a disagreeable boring, grating sensation caused by the needle as it passes through the soft parts and against the bony surfaces; but this can be diminished to some extent by the fractional instillation of novocain as the needle is introduced. Various attempts have been made to render the treatment more endurable, and it occasionally becomes necessary to resort to general anesthesia. Ether, chloroform, and nitrous oxid have been

used, but Sicard correctly condemns their habitual employment, since it is essential to have the patient's coöperation throughout the injection. The author has never found it necessary to produce general anesthesia, and, upon one occasion, made a successful injection of the gasserian ganglion in which the pain was so slight that there was no indication of the accuracy of the injection until the characteristic trigeminal anesthesia developed. Should the technic become so perfected as to guarantee accurate direction of the needle without the assistance of the patient, a general anesthetic may be more largely administered. Pollock and Potter, and Howell, from a series of fluoroscopic studies, claim to have attained this degree of accuracy.

With the injection of the alcohol, and sometimes before, if the needle is actually within the nerve trunk, there is a sharp attack of pain throughout its distribution resembling the paroxysms with which the patient is already too familiar. This is followed by immediate relief of pain, and the development of anesthesia or a sense of swelling in peripheral distribution of the injected nerve. The loss of sensation persists for several months; but a return of sensation does not necessarily indicate an immediate recurrence of the paroxysms. Tactile sensation is the first to return, then thermic, and, lastly, painful impressions.

Accidents Following Injection.—A successful injection should result in no serious consequences, and the author knows of no fatality except in Härtel's case, which was said to be due to careless technic. Minor unpleasant effects have, however, been observed, but most of them are temporary, and many may be avoided as the technic of the operator becomes more skillful. Not infrequently during the injection, or upon withdrawing the needle, hematoma has developed. In some cases it may be extensive, and discolor the skin as far as the clavicle, or completely close the eye for a week or more; but ordinarily, there is no hemorrhage or it is limited to the soft parts of the cheek or orbit, and is more likely to occur in injections of the maxillary nerve than when the third division is injected. The blood is finally completely absorbed without doing any permanent damage. Nasal or pharyngeal hemorrhage has sometimes followed too deep a penetration when injecting the second or third divisions. Care should be taken to avoid injecting the alcohol into a vessel because of the likelihood of thrombus formation, and Sicard describes such an accident with resulting gangrene. Temporary ocular palsies, facial paralysis, erysipelas, stiffness of the muscles of mastication, and sloughing of the soft parts, have been recorded among the accidents following injection; but none of them are said to be permanent. Facial paralysis and erysipelas are obviously due to errors in technic, although it is stated that erysipelas may sometimes be a mere recrudescence. Increased lacrimation, excessive nasal discharge, and loss of taste upon the anterior part of the tongue have been observed after injection of the third branch.

Duration of Relief.—Much of the adverse criticism concerning the injection of alcohol is to be attributed, then, to the painfulness of the treatment, the uncertainty of success at any one treatment, the unpleas-

ant consequences experienced by those unprepared to perform the operation, and the necessity of repeated injections. The duration of relief varies within wide limits, and depends upon the character and accuracy of the injection. The freedom from pain following a superficial injection varies from a few weeks to a month or two, although the writer has known such an injection to give relief as long as seven months. A successful deep injection is almost always followed by complete relief for a period of months or years. In the majority of cases, the treatment is effective for about eighteen months, though it is not uncommon to secure much longer intervals. One of the writer's patients was relieved for seven years, and an instance has been recorded in which the pain had not recurred after an interval of ten years. Sicard states that, in many hundred injections, he knows of only two instances in which the treatment failed, and in one of the writer's patients the result was unsatisfactory after two attempts to inject the nerve. Beckman found that in a series of cases treated by injection, neurectomy, and the ganglion operation, there was slightly longer relief after the injection than after neurectomy. The average duration following injection was 9.4 months, and the longest five and a half years. After neurectomy, the average relief was 8.4 months, and the greatest duration only two years. Payne records a case in which there was freedom from pain for three years after a deep injection. Perhaps the early recurrence in some cases may be due to incomplete injection or to the presence of an untreated dolorigenetic zone in an area adjoining that innervated by the injected nerve.

It is generally believed that a repeated neurectomy is seldom as effective as the first operation, and a similar opinion has been expressed concerning the injection of alcohol. Frazier, Peet, and others who question the value of repeated injections, maintain that alcohol, in time, loses its efficiency. The writer, during his earlier experience, was inclined to hold the same opinion. A patient, in whom the mandibular nerve was first injected in 1910, was relieved for ten months. Upon the return of pain, many attempts were made to reinject the nerve; and although the treatments were frequently followed by the characteristic sensory changes indicating a successful injection, pain was never relieved for more than a few weeks. At first, the author attributed these repeated failures to the inaccuracy of technic; but, later, began to think that the alcohol had actually lost its neurolytic property. Dr. Finney, then, resectioned the inferior dental nerve and injected the central stump with alcohol. This relieved the paroxysms for about six months, although there was no anesthesia of the lower lip. With the recurrence of the attacks, the patient again insisted upon injection, and contrary to the writer's better judgment, and with confidence largely destroyed, he was persuaded to comply with the request. Repeated attempts resulted in so many failures, but finally, an injection was made which resulted in a period of relief equal to that following the first treatment; and his earlier belief that the repeated failures were due to faulty technic and not to the diminishing effectiveness of the alcohol was reestablished. Neither before nor since has the writer encountered a similar experience,

and he is confident, from having performed many repeated injections, that if the treatments are accurately executed the results are entirely satisfactory.

Not infrequently the advice of the family physician is sought in making a choice of neurectomy or alcoholic injection; and from the evidence accumulated within the past ten years, there should be no hesitancy in accepting the injection of alcohol in preference to all other methods of peripheral operation. It has stood the test of time; the duration of relief is equal to, if not greater than, that following neurectomy; it possesses the advantage of being successfully repeated in the vast majority of instances; and it is quickly performed. General anesthesia is not necessary; the patient need not be in the hospital more than a few days; relief is certain when the nerve is accurately injected; and as the technic of the operator becomes more nearly perfect, the probability of relief at the first injection can be promised with greater assurance. If the attacks are confined to the supra-orbital nerve, neurectomy is the more desirable procedure, although almost any form of peripheral operation upon this nerve is frequently disappointing. The author has seen supra-orbital neuralgia treated by both methods, and seen both methods fail; but occasionally, relief for many months may be secured by the injection of alcohol at the supra-orbital notch.

The inevitable recurrence after all forms of peripheral operation, and the increasing confidence in the effectiveness of alcohol soon led to the hope, and final successful accomplishment, of injecting alcohol directly into the gasserian ganglion without exposure, and without a cutting operation. Subcutaneous injection of the ganglion was first performed by Taptas, and later by Harris, Grinker, Vaughn, Robinson, Dorrance, Payne, Hamill, Howell, Hirschel and others. A full description of the technic will be found in the writings of Harris, and in the subsequent publications of Härtel. Beckman and Peet are of the opinion that the operation is extremely hazardous, while Frazier cautions that "the incidence of corneal complications, apart from injuries to the abducens and oculomotor nerves is not small." This, he thinks, makes the ganglion injection prohibitive. The author's experience does not confirm this opinion; but there is no question that injection of the gasserian ganglion should never be attempted by any one who is not thoroughly trained to perform the operation.

Continued experience with this method of treatment has not contradicted the conclusions expressed by the writer in an earlier publication on this subject, and he may, therefore, be permitted to refer to them indirectly here. In a large percentage of skulls, the gasserian ganglion may be injected through the foramen ovale according to either the method devised by Harris, or the one suggested by Härtel. Alcohol destroys, permanently, those nerve-cells into which it is injected. It is not only possible, but practicable, to partially inject the ganglion, and thus avoid, in suitable cases, permanent injury to the cells whose axones innervate the cornea. In none of the author's cases did keratitis develop, although partial loss of corneal sensibility has repeatedly been obtained.

Even complete corneal insensibility is frequently transitory, and of shorter duration than the cutaneous anesthesia. The duration of relief, following injection of the ganglion, depends upon the completeness with which the ganglion has been infiltrated, and the percentage of cells which have been permanently affected. Although it is impracticable to destroy the entire ganglion at any single injection, repeated injections should insure permanent relief. The probability of regeneration of the motor fibers is greater after injection than after any surgical procedure in which the motor-root has been destroyed. The longest relief obtained in the writer's series of cases is now six years, and in only four of the thirty cases have there been recurrences. Injection of the ganglion, or deep neural injections do not preclude later surgical intervention should it become desirable.

The application of the injection method of treatment has, however, its limitations, and a word of caution should be given to those who entertain the belief that, if alcohol will relieve major neuralgia, it ought to relieve every trigeminal pain. The minor neuralgias, and particularly the psychalgias, are rarely benefited by any peripheral nerve operation, and the injection of alcohol in such cases usually results in physical discomfort and failure to relieve the pain. A striking example of such misdirected therapy was recently obtained from one of the writer's patients. A young woman with marked psychoneurotic symptoms of fifteen years' duration, experienced a dull continuous pain in the left maxillary area, following the death of a parent. The usual nasal operations, irrigations of the antrum, and intranasal injection of Meckel's ganglion were resorted to without relief. Her physician then injected the gasserian ganglion with alcohol and secured complete trigeminal anesthesia, but no alleviation of the pain. Sleep then became disturbed by dreams of the various operations she had undergone; the paroxysms were intensified, and invariably associated with a definite group of past experiences connected with the death of her parent. A "rest cure" was prescribed, and during the course of treatment there was no time, even at the height of a violent exacerbation, when the pain could not be relieved by appropriate psychotherapeutic measures. Surprisingly enough, the physician who performed the injection in this instance has since published an article advocating the injection of alcohol into the gasserian ganglion as a means of securing trigeminal anesthesia preparatory to the performance of minor nasal and pharyngeal operations! Under local anesthesia, thus induced, he removed the tonsils, performed two minor nasal operations, and evacuated an alveolar abscess. It is hoped that the procedure will not become generally adopted.*

SURGICAL TREATMENT.—The only surgical procedure worthy of consideration is the removal of the gasserian ganglion, or avulsion of its sensory root. Other surgical measures, such as nerve stretching, neu-

* The author referred to in the above paragraph has since contributed a second paper upon injection of alcohol into the gasserian ganglion in which an extensive use of the operation is recommended.

rectomy, and neurotomy, have been almost entirely superseded by alcohol injection; and ligation of the common carotid artery and general blood-letting are of historical interest only. Rosenthal has recently reported seven cases of trifacial neuralgia and other head pains successfully treated by removal of the appendix.

CHOICE OF A METHOD OF TREATMENT.—Except as a temporary, palliative measure, or until other means may be procured, treatment by drugs, electricity, and local applications is generally useless. A decision must then be made between injection with alcohol and the major surgical operation. Selection of the major operation is influenced, to some extent, by the surgical inclination of the physician, the greater assurance of permanent relief from the radical operation, the skill of the available surgeon, and the clinical conditions of the individual case. If injection is decided upon it must then be determined whether it shall be superficial, deep, or ganglionic.

Since the majority of patients affected with the disease are within the fourth or fifth decade, age alone does not ordinarily enter into the choice of the measure to be adopted. In the younger patients, however, who have the prospect of a longer life with repeated recurrences, especially if more than one branch is involved, it is advisable to resort to the major operation; but injection of the ganglion may be recommended, provided the mandibular nerve is one of the affected branches. If, however, the disease is confined to the first and second divisions, the major operation should unhesitatingly be advised; since, under these conditions any attempt to inject the ganglion must necessarily expose the uninvolved mandibular nerve to a certain amount of damage. There is also a small percentage of skulls in which the foramen ovale cannot be entered, and failure to inject the ganglion would not only result in failure to relieve the pain, but might initiate paroxysms in the unaffected third division. Should the patient refuse the major operation, a deep injection of the second branch and a superficial injection of the supra-orbital nerve may be made.

In those of advanced years the situation is different. If there are bodily infirmities which increase the risk of the major operation, injection of the ganglion, in suitable cases, should be the method of choice; although such a patient can usually be kept fairly comfortable throughout his remaining life by deep nerve injections alone. Age and infirmity, in the author's experience, have been no contraindication to treatment by injection, and the author recalls a patient, 79 years of age, who had marked arteriosclerosis, high blood-pressure, hypertrophic cirrhosis of the liver, aortic insufficiency, and chronic interstitial nephritis upon whom a successful injection was practiced without the least unpleasant consequences.

Every patient should be given the privilege of choosing an injection before resorting to more radical measures, for it in no way precludes the later adoption of the major operation. The author's custom has been to state frankly to the patient, at the outset, the four methods of treatment: drugs, deep injection of alcohol, alcoholic injection of the

ganglion, and the intracranial operation. An explanation of the probable results to be expected from each method is then made, and the patient is allowed to make his choice.

In the bilateral affection, injection of the ganglion has distinct advantages over all other methods. In performing the major operation, the motor root is not infrequently included in the avulsion; and unless every assurance can be given that the motor fibers will not be damaged, bilateral avulsion is inadvisable if not impracticable. It might be suggested that the operation be performed in two stages, and if there is no indication of injury to the motor root at the first operation, the opposite side may then be treated. If, however, the motor root has been torn, regeneration of its fibers is, under these conditions, extremely improbable and precludes the possibility of a second operation for the untreated side. Injection of the ganglion seems to obviate this difficulty. Although the motor fibers may be destroyed by the injection, the neurilemma remains intact; the motor nucleus is not affected, and regeneration is almost certain within six or eight months. During the period of regeneration, a deep injection of the second division and superficial injections of the first and third divisions may be made upon the opposite side; and upon recovery of motor function to the side first injected, the opposite ganglion may be treated without risk of complete motor disability.

Prognosis.—Trigeminal neuralgia is a chronic disease; it is not self-limited and shows no tendency to spontaneous recovery, although Dana is of the opinion that the natural duration of the affection is from 6 to 12 years. Remissions of variable duration are, however, not uncommon; but the paroxysms generally increase in intensity with advancing years. It is never fatal, and in no way shortens life. Suicide occasionally terminates the affection, and Fothergill states, with some skepticism, that Dr. Lawrence Bausch (1665), President of the *Societas naturæ curiosorum*, is said to have died of the disease.

Pathology.—No uniform, constant lesion has been described within the ganglion or its branches, which serves to establish a pathological basis for the disease. The peripheral nerves, ganglion, sensory root, and motor nuclei have been studied by many competent observers, who have found various changes within the cells and fibers, but it is difficult to form an opinion as to the primary seat of the changes which have been described. Occasionally, the histological structure of the nerve and ganglion is said to be quite normal. Frazier is of the opinion that the lesion is essentially a sclerotic process within the ganglion, although it is more generally believed that the disease is the result of a primary ascending neuritis. Spiller, in one of his earlier studies, expressed this view, and stated that he has seen two instances in which the affection was thought to be secondary to diphtheria and erysipelas. Dana, Putnam, and Thoma have described thickening of the arteries, both within the nerve trunk and within the ganglion, and have attached special importance to this pathological finding. The interpretation of pathological changes is, however, difficult because of the in-

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frequency with which ganglia are obtained from patients who have not previously been subjected to some form of peripheral operation. Such studies have, however, been made by Spiller, who found some sclerosis of the intraganglionic vessels, and by Schwab, who observed marked cellular disintegration.

The peripheral nerves have been examined by Dana, Putnam, Spiller, Thoma, Krause, and Mears, who have observed swelling of the axis cylinders and myelin sheath, slight round-cell infiltration, accumulation of myelin droplets by the osmic acid method, increase in connective



FIG. 1.—CROSS-SECTION OF THE MANDIBULAR NERVE AND ITS ACCOMPANYING BLOOD-VESSEL IN CASE OF TRIGEMINAL NEURALGIA.

Chronic arteritis. Thickening and disintegration of the intima with involvement of the media. (Elastic tissue stain.) ($\times 100$.)

tissue, and obliterating endarteritis. The cause of these changes could not be determined; but since they were, in some cases, more intense at the periphery than about the ganglion, it was believed that the process was an ascending parenchymatous neuritis. The author examined a portion of the mandibular nerve, removed by Dr. Cushing, from a woman, 59 years of age, who had been subject to neuralgia in this division for three years. There was no special thickening of the palpable arteries, but the systolic blood-pressure varied from 180 to 220 mm. Hg. The sections showed diffuse thickening of the vessels, obliterating endarteritis, and slight swelling of the axis cylinders; but there was no evidence of acute or chronic inflammation (Figs. 1 and 2).

The gasserian ganglion examined by Head was so perfect that he

made use of it in demonstrating normal nerve-cells; while others have described changes, within the ganglion, varying from the most insignificant chromatolysis to complete cellular disintegration and degeneration of the intraganglionic fibers. Beckman studied six ganglia, removed at the Mayo clinic, and all showed marked evidences of inflammation. Spiller has recorded the changes observed in two ganglia removed by Dr. Cushing and seven from Dr. Keen's clinic. Some of the specimens were only slightly altered; but most of them showed thickening of the blood-vessels, cellular changes, swelling of the axis cylinders and medullary sheath, intense degeneration at the periphery of

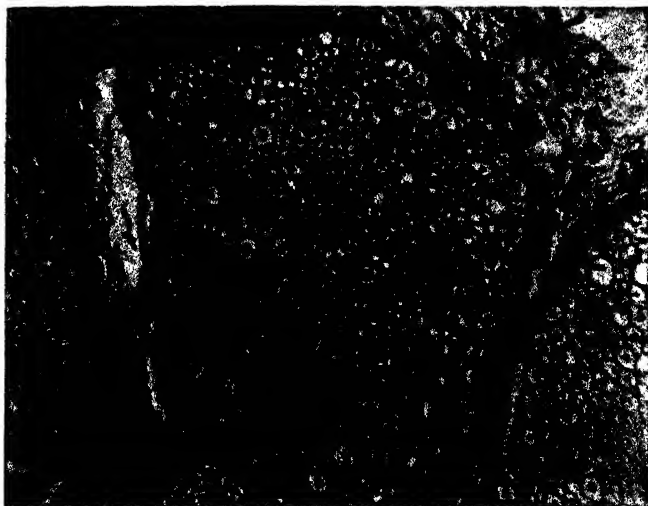


FIG. 2.—CROSS-SECTION, MANDIBULAR NERVE IN CASE OF TRIGEMINAL NEURALGIA.

Except for occasional swelling of the axis-cylinders, the nerve appears to be quite normal. (Hemalum-acid-fuchsin stain.) ($\times 300$.)

the ganglion by the Marchi method, and concentric deposits within the ganglionic substance. In one of the ganglia, the cell changes were intense, and in another there was almost complete vascular occlusion. Rose has made similar observations, and Schwab attaches special importance to perinuclear pigmentation and interstitial sclerosis; but in none of his four ganglia did he find marked vascular changes. McCarthy has observed pronounced cellular disintegration, and Barker found "scarcely a normal nerve-cell" in one of the two ganglia he studied, but there was no sclerosis, and only moderate thickening of the blood-vessels. Krause describes sclerosis and cellular changes in seven specimens, and in one instance, degeneration of the sensory root; although Spiller, in a similar study, found the sensory root normal.

In examining fifteen ganglia, removed by Dr. Cushing, the author

did not find a single instance in which the ganglion was perfectly normal. All of them showed variable degrees of cellular disintegration, although not all of the cells in any one ganglion were equally affected. Some of the more pronounced cellular changes are illustrated in the photomicrograph (Fig. 3). In several specimens there were evidences of general sclerosis, and in all but two ganglia the pericellular nuclei were increased. Occasionally, the capsular cells appeared to have invaded the entire ganglion and filled the spaces left vacant as the ganglion cells disintegrated. Many concentric deposits were present in



FIG. 3.—SECTION OF THE GASSERIAN GANGLION IN CASE OF TRIGEMINAL NEURALGIA.

The cells are retracted from their membrane, irregular in contour, and show increased pigmentation, excentric nuclei, chromatolysis, and vacuolization. (Van Gieson stain.) ($\times 300$.)

all of the specimens. These bodies were more frequently observed within the intraganglionic fibers than among the groups of nerve-cells (Figs. 4 and 5).

In none of the ganglia or their peripheral divisions, including the sensory root, did the nerve fibers take the myelin stain well, although there was no real degeneration. The intraganglionic blood-vessels were not especially thickened, but it was invariably observed that the vessels within the nerve trunks showed sclerosis in both the intima and the media; and these changes were more pronounced toward the distal portion of the nerve trunks.

SUMMARY.—Although no definite pathology has been established for the disease, many of the ganglia and peripheral nerves have shown distinct histological changes. The writer's studies confirm the observa-

tion of Dana and Putnam concerning the frequency of local arteriosclerosis, and its predominance within the peripheral branches. These observations together with the therapeutic evidence furnished by the relief occasionally secured from the use of those drugs which reduce vascular tone, such as large doses of strychnin, aconite, and the nitrites, are confirmatory evidence of the probable vascular nature of the affection.

The occasional association of migraine and Raynaud's disease with trigeminal neuralgia offers interesting speculation. These associated conditions are thought to depend upon a localized vascular spasm in



FIG. 4.—GASSERIAN GANGLION IN CASE OF TRIGEMINAL NEURALGIA.

The capsular cells are increased and occupy some of the ganglion-cell spaces. The nerve-cells are shrunken, and the darker cell, within the center, shows perinuclear pigmentation. (Van Gieson stain.) ($\times 100$.)

which arterial thickening may, or may not, be a factor. It is conceivable that the paroxysmal attacks in trigeminal neuralgia may be due to local angiospasms, with temporary ischemia of the ganglion, or more probably of the nerve trunk. Arteriosclerosis is said to increase the irritability of the vessel wall, and the exciting cause of the presumed arterial constriction in major neuralgia may be one of the various forms of peripheral stimuli from the trigeminal area, or the spasm may be part of a general cerebral or somatic vasoconstrictor impulse. Those who hold to the toxic theory of the disease need not find in this reflection anything to discredit their convictions, for it has been demonstrated by physiological experiments that even slight local injury to the arterial wall renders that portion of the vessel more irritable, and that it may

exhibit local constriction from the irritating effects of a circulating toxin.

The vascular pathology of the disease is opposed by those who argue that the changes which have been found in the blood-vessels are secondary to the pain; that repeated painful experiences, in time, produce arterial thickening; that the arterial spasm is not demonstrable; and that specimens of nerve and ganglion have been studied in which the vessels are normal. But arterial changes are not uniformly demonstrable in Raynaud's disease, in intermittent claudication, in mi-

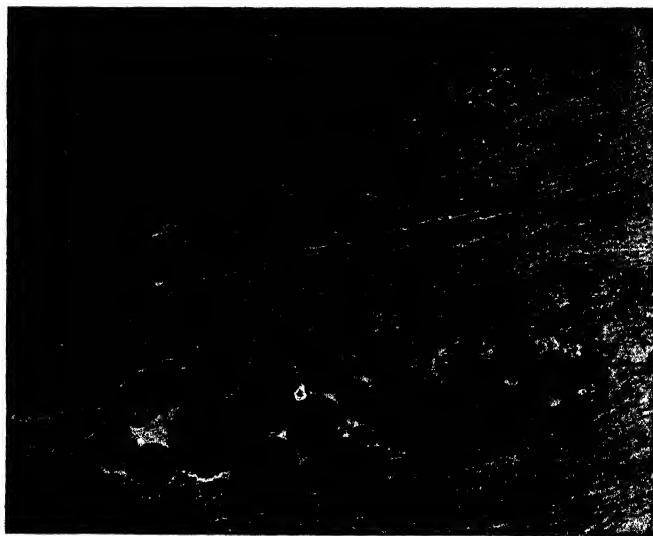


FIG. 5.—GASSERIAN GANGLION IN CASE OF TRIGEMINAL NEURALGIA.

Upper part of photograph shows retracted nerve-cells and proliferation of capsular cells; in the lower part, the intraganglionic fibers with many concentric deposits are exhibited. (Van Gieson stain.) ($\times 50$.)

graine and other diseases attributed to arterial spasm. In a specimen removed from a patient of the writer who had suffered from neuralgia for only six months, the arterial changes were equal to those observed in some specimens obtained from patients who had had the disease for several years. Thoma studied the supra-orbital nerve of a patient who suffered from neuralgia of this nerve and general arteriosclerosis. The vessels about the nerve were thickened to a marked degree, and were regarded as the primary lesion, while the general arteriosclerosis was attributed to the repeated and long-continued suffering. Diffuse arteriosclerosis is, however, no indication that the vessels within the nerve trunks or ganglion are similarly involved. The writer studied the ganglion and its branches, removed at autopsy, from a patient 79

years of age who had never suffered from trigeminal neuralgia. The systemic and peripheral arteries showed intense thickening and atheromatous patches; but the vessels of both gasserian ganglia and their peripheral branches showed no marked changes. The cells in both ganglia were slightly altered in shape and internal structure (Fig. 6).

Historical Summary.—The gasserian ganglion was so named by Antonius Raymond Balthazar Hirsch in honor of his preceptor, John Laurentius Gasser, an anatomist of the eighteenth century.

The disease, with which the ganglion and its branches are affected,



FIG. 6.—GASSERIAN GANGLION IN MALE, 79 YEARS OF AGE, WHO HAD NEVER SUFFERED FROM TRIGEMINAL NEURALGIA (AUTOPSY).

Vessels of ganglion and its branches not specially thickened. Nerve-cells shrunken. (Van Gieson stain.) ($\times 50$.)

was first systematically described in 1776, by Dr. J. Fothergill, an English physician, and designated by him as: "A Painful Affection of the Face." It had probably existed before Fothergill's time, but had been mistaken for gouty and rheumatic affections, or confused with the various types of facial spasm. The younger Fothergill (S.), a nephew of the former, published in 1804 an excellent historical review of the disease, in which it is stated that the affection was known as early as 1665, when it was claimed that Dr. Bausch died from its effects. Much of historical interest is also to be found in a paper by Pujol which appeared several years after Fothergill's original description. From the time of Hippocrates up to the latter part of the eighteenth century the disease had been confused with the several types

of involuntary laughter. The spasmodic facial contractions sometimes accompanying trigeminal neuralgia were, no doubt, responsible for the earlier designation of a certain number of cases of ris sardonique, and the raptus caninus of Aurelianus as genuine neuralgia; and Avicenna added to the confusion when he described a type of involuntary laughter associated with pain; but there is no proof that any of these conditions were real attacks of major neuralgia. The younger Fothergill, however, succeeded in collecting 52 well-recognized cases from the literature at the time his paper was published.

From the earliest writing up to the present time no entirely satisfactory name has been suggested for the disease. *Dolor faciei* Fothergilli was used by the Germans, *trismus dolorificus* by Savage, *spasmus cynicus*, *hemisrania idiopathica*, *la grande nevralgie* by Lévy, and *tic douloureux* by André and Thouret; but the objection to each of these terms is apparent. The disorder does not resemble hemisrania; it is not associated with definite involuntary spasm, nor is it a form of involuntary laughter; and *tic douloureux* may be a painful spasm in any part of the body. The younger Fothergill objects to attaching his uncle's name to the disorder, and prefers to speak of it as *faciei morbus nervorum crucians*. It has also been called essential neuralgia of the fifth nerve, neuralgia quinti major, surgical neuralgia, epileptiform neuralgia, *tic epileptiforme*, trigeminal neuralgia, and facial neuralgia. The last is particularly objectionable because of the suggested involvement of the seventh nerve; and "trigeminal neuralgia," unless qualified, includes the minor as well as the major neuralgias of the fifth nerve.

It seems that the objection to perpetuating personal names in the designation of diseases is generally approved, but unless a more suitable term can be selected the objection should not be too forcibly maintained. It is true that most writers upon the subject use the term "trigeminal neuralgia" to designate the affection described by Fothergill; but in the every-day communications between physicians there are many who either have no conception of the disease which Fothergill described, or else regard every pain in the face as trigeminal neuralgia. No apology need be made, then, if it is stated that the affection which has been described in these pages is none other than the definite clinical entity so admirably presented by the English physician whose name deserves to be perpetuated in—"Fothergill's disease."

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CHAPTER XI

CEREBRAL PALSIES OF CHILDHOOD

BY CHARLES S. POTTS, M.D.

Etiology, p. 91—Symptomatology, p. 91—Pathology, p. 95—Diagnosis, p. 96—Treatment, p. 99—Prognosis, p. 100—Historical summary, p. 101.

When speaking of the cerebral palsies of children, that which is usually meant is paralysis of more or less extent due to abnormal birth or occurring as a sequel to one of the infectious diseases. It must be remembered that there are other cerebral lesions causing paralysis in childhood. These will be mentioned in the discussion of differential diagnosis.

Etiology.—The fuller etiology of the two main divisions will be given when discussing symptoms and pathology (q.v.). In regard to those cases of cerebral palsy due to injury at birth, however, the question has arisen whether it is more liable to be caused by a prolonged labor or by use of the forceps. This, of course, has a bearing on prophylaxis. In the statistics quoted on p. 93 it will be noticed that more cases were attributed to prolonged labor than to use of the forceps. Sachs, discussing this subject in his book on the "Nervous Diseases of Children," states that tedious labor is much more dangerous than the forceps and that most cases occur in first children. Sharpe and Farrell state that prolonged labor should be avoided whenever possible. They also state that the cesarean operation is less dangerous to the child than the high forceps. This, however, is largely a question for the obstetrician.

Symptomatology.—CEREBRAL PALSIES DUE TO ABNORMAL BIRTH.—These, also known as Little's disease, may be due either to injury during birth, to premature birth, or to cerebral disease occurring during intra-uterine life.

Injury during Birth.—Recognition of those injuries present just after birth is very important. These depend upon whether the lesion (hemorrhage) is supratentorial or infratentorial. If the former, the child may be born asphyxiated, or cyanosis may not appear until several days later. In addition the child will be restless and cross, will refuse to nurse, and within a few days have convulsions, which may be confined to one side. Examination will show a tense or bulging anterior fontanel, associated with some of the following symptoms: Muscular rigidity, edema of the eyelids, subconjunctival hemorrhage, irregularity of the pupils, more or less papillo-edema, and a slow pulse. If the latter, there will also be asphyxia, which, according to Seitz, may not be marked

until after several days, when the nerves at the base of the brain having to do with respiration become involved. At this time also there will be irregular and labored breathing. The child will be apathetic. Tension of the fontanel is not usually so great, at least during the first few days, as when the hemorrhage is supratentorial. Muscular rigidity and retraction of the head may be present. In the presence of any combination of the above symptoms lumbar puncture should be done, when if hemorrhage has occurred a bloody cerebrospinal fluid under increased pressure will be obtained. Asphyxia and convulsions occurring within a few days after birth are an indication for the immediate performance of this procedure.

In many instances, the cause of the trouble is not recognized at this time and if the child survives, nothing wrong may be noticed until it is old enough to sit up. It will then be observed that it is unable to do so, and if supported in the sitting position, the head will fall to one side or the other owing to the weakness of the muscles of the neck. In other cases, the occurrence of either epileptiform convulsions or spasmodic movements of the arms or legs will be the first symptoms to attract attention.

Examination at this time, or later, will usually determine the existence of weakness and rigidity of either the arm and leg of one side (hemiplegia), of both legs or both arms (paraplegia), or of both arms and both legs (diplegia). Rarely these symptoms may be confined to one limb (monoplegia). The tendon jerks (Achilles', knee, biceps and triceps) of the affected limbs will be increased and ankle clonus is often present. In cases in which the spasticity is very great, these reflexes may apparently be diminished. This is due to the fact that the muscles are so hypertonic that they are unable to contract. The Babinski reflex will be present. In children under two years old this has no significance as it is then normal. In the course of some months deformity occurs in the affected limbs due to the over-action of the flexor muscles (contractures) and in time the joints become more or less ankylosed. If the patient can walk the gait is more or less spastic. Sometimes the overaction of the adductors is so great as to cause the legs to be crossed (cross-legged progression). In this connection it must be remembered that in some instances actual weakness is not marked and the disability is due to the intense spasticity. If one side only is affected, the gait is of the hemiplegic type. The muscles will respond normally to the electric current. The limbs are often cold and cyanotic. In many of these cases there are present involuntary, slow, vermicular movements, especially of the hands and fingers and sometimes of the feet, toes and facial muscles. These are known as athetoid movements and the condition as athetosis. Sometimes these movements more closely resemble those of chorea. Clonic spasms of the arm muscles may also occur. If, as is sometimes the case, there has been considerable return of power and very little muscular rigidity, these movements may be mistaken for those of Sydenham's chorea.

Anarthria is sometimes present due to involvement of the muscles

of the tongue and larynx, and there may be paralysis of the muscles of the face. The latter will be of the supranuclear type, i.e., only the muscles about the angle of the mouth will be affected. Weakness of the ocular muscles and nystagmus may be observed. Aphasia is exceedingly rare, and if it occurs is usually soon recovered from. A very serious feature is the frequent occurrence of mental impairment and epileptiform convulsions, which, if the lesion is unilateral, may be jacksonian in type. Spratling attributes 11 per cent. of 1,070 cases of epilepsy to cerebral hemorrhage at birth or shortly after. Of 388 cases analyzed by Turner, 5.9 per cent. were due to this cause.

An analysis of 5,430 cases of varying degrees of mental impairment made by Beach, Shuttleworth and Barr showed that 9.74 per cent. were attributed to pressure due to prolonged labor and 2.37 per cent. to the use of the forceps.

The foregoing description is that of the type usually met with; there are, however, other types which are seen much less frequently. Cases have been described in which there is diplegia with marked *hypotonia* of the muscles. The motility of the limbs may be normal. If the child becomes able to walk the gait resembles that of cerebellar disease and there is incoördination of the arms and hands. There is no muscular atrophy, the electrical reactions are normal, and the kneejerks may either be increased, diminished or lost. Speech defect is usually marked. This has been termed the *atonic-astasic type of infantile cerebral paralysis*, or *infantile cerebrocerebellar diplegia*.

Another group has been termed *cerebellar diplegia*. In these cases there is ataxia of the cerebellar type, with nystagmus, and slow, jerky and monotonous speech. The mentality is good. Still another group has been described in which the prominent symptom is incoördination. This has been termed *diataxia cerebialis infantilis* or the *ataxic type of cerebral birth palsy*. These are characterized by ataxia, bilateral in distribution, which involves gait, station, the use of the arms and legs and the muscles having to do with speech. The incoördination is also present when in the recumbent position. There is neither paralysis nor spasticity. The tendon reflexes are present but not increased, and the Babinski reflex may be absent. Epilepsy and mental defect were not observed in the cases described.

Premature Birth.—The clinical history of these patients is similar to that of those above described, excepting that the child was born prematurely and the early symptoms (asphyxia, convulsions, etc.) did not occur. Epilepsy does not develop so frequently as in those due to trauma at birth.

Cerebral Disease Occurring during Intra-uterine Life.—A certain percentage of cerebral palsies in children are due to syphilis in one or both parents. In such stigmata of hereditary syphilis will probably be found and the Wassermann test will likely be positive in the affected parent. One of the febrile infectious diseases occurring in the mother, or trauma, especially abdominal, sustained by her while pregnant, may also cause an encephalitis in the child and resulting paralysis. In this

connection may be mentioned severe jaundice (*icterus neonatorum*) occurring in the newborn child. The history in these cases as regards the discovery and development of the symptoms will be similar to that of those resulting from premature birth, excepting that the child was born at full term and either evidences of syphilis in the parents, the occurrence of an acute infectious disease or trauma during pregnancy, or severe jaundice was present in the patient within a few days after birth. In one of the cases of jaundice described by Spiller there was also a history of premature birth.

CEREBRAL PALSIES OCCURRING AS SEQUELÆ OF ACUTE INFECTIOUS DISEASES.—While it is well known that *cerebral apoplexy* is of common occurrence after fifty years of age, it does not seem to be generally known that it is exceedingly common during the first decade of life. The following statistics collected by Thomas will make this clear:

STATISTICS OF APOPLEXY, SHOWING DECADE DURING WHICH APOPLECTIC ATTACK OCCURRED (Thomas)

Decade	Males	Females	Total
First.....	58	77	135
Second.....	16	9	25
Third.....	43	19	62
Fourth.....	73	29	102
Fifth.....	88	49	137
Sixth.....	99	44	143
Seventh.....	86	19	105
Eighth.....	20	5	25
Ninth.....	6	0	6
	489	251	740

The death rate from apoplexy is greater during the first ten years of life than at any other period up to fifty. The reason for this is the fact that any of the acute febrile diseases may cause degeneration of the blood-vessels and a resulting apoplexy. Some of the cases occurring during this period may also be due to *hereditary syphilis* causing disease of the blood-vessels; others may be inflammatory.

The usual history of these patients is, first, the occurrence of diphtheria, scarlatina, measles, whooping-cough or some other infectious disease. Either during the progress of the disease or while convalescent, the child is seized with convulsions and is unconscious for a varying period of time. In some instances this may be preceded by evidences of headache and muscular rigidity, and retraction of the head may be present. When these latter symptoms occur the lesion is probably inflammatory (encephalitis) and not apoplectic. If the child survives, after consciousness returns, it will usually be found to be hemiplegic. When the lesion is left-sided, aphasia may be present but usually soon disappears. In left-handed children a right-sided lesion will so act. In these cases hemiplegia is the usual result, while in those due to abnormal birth, diplegia or paraplegia is more common. This paralysis presents

the characteristics described on page 92. In addition, if due to encephalitis, paralysis of cranial nerves due to involvement of their nuclei, and sometimes ataxia, due to involvement of the parietal lobes or cerebellum, may be present. As the patient grows the paralyzed side may not develop to the same extent as the normal, the limbs may be shorter and the muscles smaller. Epilepsy, imbecility and idiocy are frequent results of these lesions.

Pathology.—**CEREBRAL PALSIES DUE TO ABNORMAL BIRTH.**—In the cases due to injury at birth, the immediate result is *hemorrhage*. This is usually either subdural or intrameningeal. In locality, it may be either at the convexity (the usual location in those who survive), or the base. Less frequently it may be in the brain substance or within the ventricles. Meningeal hemorrhage, if over the convexity, is caused by the overlapping of the parietal bones which occurs during the process of molding the fetal head, which tears the meningeal veins just before they enter the superior longitudinal sinus. The hemorrhage with the resulting clot is usually most pronounced over the motor region, which accounts for the usual occurrence of spastic diplegia or paraplegia. If it involves the parietal area, the ataxic type will result (Hunt); if the frontal lobes, the astasic-atonie type. Thrombosis in the superior longitudinal sinus may also be found. More or less softening of the cerebral cortex as a result of the interference with the circulation occurs, which results in degeneration of cortical nerve cells.

Hemorrhage involving the base of the brain (infratentorial) is caused by similar tearing of the veins entering the lateral sinuses which at times may also be ruptured. It is possible that the cerebellar type may be due to such a lesion.

The lesions which may be found later in life are: (1) Porencephalus, meaning a cavity in the brain cortex communicating with the arachnoid spaces and penetrating deeply into the brain, even into the ventricles. (2) Diminution in the size of the convolutions (microgyria). (3) Unilateral or bilateral atrophy of the brain due to diffuse sclerosis. (4) Localized areas of sclerosis (tuberous sclerosis). (5) Microcephalus. (6) Cysts either in the cortex or meninges, which may have hemorrhagic contents. These lesions may be found in the brain cortex, in the white matter of the brain and in the basal ganglia and their neighborhood. Porencephalus may be a true malformation, but is frequently an end result of the processes above described. In these cases it is usually found in the area supplied by the middle cerebral artery, i.e., the motor region. A number of autopsies have been made in which no gross lesions were found. This is especially so in cases due to premature birth. In these cases, however, there have been found microscopically a sparseness, a fineness and a lack of development of the fibers constituting the pyramidal tracts. Undeveloped cells in the brain cortex have also been found, and the evidences of lack of growth of the brain itself above described are frequent.

In the cases due to disease or trauma sustained by the mother during the intra-uterine life of the child, the conditions found will depend upon

the cause. In many instances nothing characteristic will be observed, the appearances resembling the terminal lesions found after a meningeal hemorrhage. If due to hereditary syphilis, disease of the blood-vessels (endarteritis), gummatus infiltration and chronic inflammation of the meninges or isolated gummata may be found if death occurs early. When due to infectious disease in the mother, as typhoid fever, or to a trauma sustained by the mother, evidences of encephalitis may be present. According to Orr and Rows, in those due to a febrile infectious disease a toxic, hyaline thrombosis of capillaries and perhaps of larger vessels will be found. Two cases have been described by Déjerine (quoted by Spiller) in which the symptoms resembled a diplegia due to a cerebral lesion, but were found to be due solely to one of the spinal cord, which was myelitic and had occurred *in utero*.

CEREBRAL PALSIES FOLLOWING INFECTIOUS DISEASES.—The lesions found early when the condition has occurred as a sequela of one of the febrile diseases of childhood are frequently those of apoplexy, i.e., either hemorrhage, or thrombosis or embolism in one of the cerebral arteries, usually the middle meningeal or one of its branches. To have either hemorrhage or thrombosis, disease of the artery is essential, and this is usually a fatty degeneration as contrasted with endarteritis and atheroma which precede apoplexy in the adult. Many of these cases are, however, not apoplectic in origin, but are due to inflammation of the cortical cells and often those of the cranial nerve nuclei and cerebellum as well (encephalitis, poli-encephalitis). The terminal changes, whether due to apoplexy or inflammation, are similar to those caused by trauma at birth described above, excepting that encephalitis does not cause porencephalus. Dana and Gere have described a case of a child who from the age of four months suffered from continual choreic and tonic spasms without motor paralysis which apparently followed an attack of encephalitis. Evidences of degeneration of cells of the motor cortex, especially of their dendrites and fibrils, were found. There was no lesion of the pyramidal tracts, excepting a thinning of the fibers.

Diagnosis.—While *asphyxia with cyanosis of the head and convulsions followed by coma*, occurring in a recently born child, are very significant of meningeal hemorrhage, it must be borne in mind that asphyxia itself after birth may be due to other causes. The most important causes are disturbance of the placental circulation, interference with the cord, as undue pressure, feebleness of the child, premature birth, malformations of the respiratory or circulatory organs, disease of the lungs, as atelectasis and pneumonia, syphilis, compression of the air passages by large glands and pleural exudates. When it occurs, therefore, with none of the symptoms detailed on page 91 present, it is probably due to one of these causes. Later *cerebral birth palsies* may have to be distinguished from *injury to the brachial plexus* occurring during labor, the so-called obstetric paralysis or birth palsy of Duchenne. In this the disability will be confined to one arm, which will be flaccid. If examined early there will probably be swelling and tenderness over the brachial

plexus in the neck. Later wasting of the muscles will occur, and they either will not respond to a faradic current, or will require a stronger one than do the normal muscles. The greatest weakness will be in the muscles about the shoulder joint, while in cerebral palsy the distal muscles are usually most affected.

Athetoid and choreiform movements, when present in cases in which marked improvement in weakness and rigidity has occurred, have been mistaken for *Sydenham's chorea*. Attention to the history of the early appearance of the symptoms, with the probability of some slight evidence of paralysis or spasticity being found by careful examination, should prevent error (p. 92). It must also be borne in mind that there may be motor weakness in true chorea (paralytic chorea), but this develops acutely some time after birth and no evidence of lesions of the pyramidal tracts will be found.

Multiple sclerosis occurring in early life may present somewhat similar symptoms. This, however, does not develop until several years after birth. Atrophy of the temporal halves of the optic disks is often present and intention tremor, scanning speech, and nystagmus are marked in typical cases.

Hydrocephalus is a cause of spastic paralysis developing in early life. It may also be a cause of difficult labor and asphyxia. The enlarged and peculiar shape of the head will distinguish the condition from hemorrhage.

A group of cases to which various titles have been applied but which may be generally spoken of as *family spastic paraplegia*, may be confounded with the cerebral palsies above described. This may be especially so in those which at times follow one of the febrile infectious diseases of children. They are family diseases affecting various members of the same generation and different generations. Isolated cases, however, frequently occur. The symptoms may appear any time from the second or third year to middle life. Usually the child develops normally until this time. Cases have been reported, however, in which the patient learned to walk late and mental development was retarded from the beginning. Such may present difficulty in diagnosis unless other members of the family are affected or atrophy of the optic nerve occurs, which, of course, is not characteristic of cerebral palsy due to the causes above mentioned. In most of these cases the lesions are spinal, in others a diffuse cerebral sclerosis has been observed (Pelizaeus-Merzbacher's disease). Rhein has classified them in seven groups, according to the symptoms observed:

1. Spastic paralysis of the legs.
2. Involvement also of the arms with or without mental failure.
3. Symptoms indicating involvement of the cerebellum. Spastic paralysis with cerebellar symptoms, nystagmus, scanning speech, with or without mental deficiency.
4. Bulbar symptoms added to the spasticity which involves the arms and legs.

5. Spastic paraplegia associated with muscular atrophy, either in the arms or legs.

6. Tremor in the arms or legs or both may be associated with spastic paraplegia. With these cases may also be classified cases in which the symptoms are those of disseminated sclerosis or similar to these (Pelizaes-Merzbacher's cases of aplasia axialis extracorticalis congenita and Krabbe's cases).

7. Family spastic diplegia.

In addition to these Purves Stewart has reported a family of which a number of members very early in life developed optic atrophy with spastic paraplegia and ataxia.

Progressive lenticular degeneration (Wilson's disease) may be confounded with cerebral palsy of childhood, if attention is not paid to the fact that it did not develop until several years after birth.

Rickets may cause muscular rigidity and spasmodic seizures. They are apt to be transient. Other signs of that disease will be present and symptoms characteristic of involvement of the pyramidal tracts will be absent.

The *atonic-astatic type* must be distinguished from *amaurotic family idiocy* (Tay-Sachs' disease) and *amyotonia congenita* (myotonia congenita). Both of these appear after the child is several months old. In the former there will be failure of vision with the appearance of the optic nerve peculiar to the disease. It is a family disease and most frequently is found in Jews. In the latter the tendon jerks are absent and if more than two years old the Babinski reflex is absent. There is no failure in mental power. It is true that sometimes in the cerebral palsy the knee-jerks and Babinski reflex will be absent, but it is not usual, and the symptoms will be noticed earlier, as a rule, than those of amyotonia congenita.

Cases of poliomyelitis have been reported as occurring *in utero*. There is a possibility that these may be mistaken for cerebral palsies. In the former the paralysis is not likely to be hemiplegic or diplegic in distribution, but will probably involve a group or groups of muscles. These will be flaccid, atrophied, with a more or less developed reaction of degeneration, the tendon reflexes controlled by affected muscles and the Babinski reflex will be absent.

The *ataxic type* may resemble somewhat *hereditary ataxia*, either the *Friedreich* or *cerebellar type*. The former disease usually does not develop until about the fourteenth or fifteenth year. It is a family disease. The knee-jerks are absent, and nystagmus is present. In the cerebellar type optic atrophy will also be present. This type may also simulate *cerebellar disease* as tumor, but this will in most cases develop after the child is several years old and if a tumor, papilledema will probably be found.

Spina bifida occulta has been a cause of weakness and ataxia of either arms or legs. This, when present, is not spastic and an x-ray examination will reveal the deformity.

The group of cases following an infectious disease (p. 96) may be mistaken for *acute anterior poliomyelitis*. The history of onset will be different, and the clinical points of difference have been mentioned on page 98.

Multiple neuritis is a comparatively frequent cause of paralysis following infectious disease. In this the muscles are flaccid and atrophied, the tendon reflexes of the affected limbs are lost, the Babinski reflex is absent. There will be tenderness over the affected nerve trunks and more or less pain. An exception to this is neuritis following diphtheria in which pain and tenderness are usually absent. In this, however, as in neuritis due to other causes, the onset is more or less gradual, and not apoplectic; consciousness is preserved; and convulsions do not occur.

Brain tumor affecting the motor centers or tract may cause *hemiplegia*. This, however, develops gradually and is usually associated with jacksonian attacks, headache and other signs of tumor.

Encephalitis following an infectious disease must be distinguished from *meningitis*. In the latter muscular rigidity, and retraction of the head and headache are usually more pronounced. The cerebrospinal fluid will be cloudy, contain many cells, and will not respond to the tests for sugar. Paralysis will not be so pronounced.

Treatment.—If *meningeal hemorrhage* is recognized sufficiently early, two procedures hold forth some hope of relief. **Lumbar puncture**, which has been mentioned as a diagnostic aid, also at times may be a useful therapeutic procedure. If signs of undue tension of the fontanel reappear, it should be repeated. This may be done daily for several days if necessary. In supratentorial hemorrhage, Cushing has **opened the skull and removed the clots** with success in four out of nine cases. The operation must be done within two weeks after birth. While the percentage of success is not great, it must be remembered that while some of these cases may lead comparatively comfortable and useful lives, most of them are doomed to great disability and suffering, frequently becoming charges on the community. Therefore, anything which holds out hope of preventing this is worth while.

In *apoplexy* occurring as a sequela of an infectious febrile disease, the treatment is more or less symptomatic. If the action of the heart is weak, **stimulants** may be given **hypodermically** if necessary and the **head should be low**. **Quietness** is essential and the patient should **lie on the side** as much as possible. When the child is able to swallow, **liquid diet** should be given and the **mouth and nasal passages kept clean**. If *encephalitis* is believed to be the lesion, treatment appropriate for that condition should be employed.

Later in life Sharpe and Farrell have performed **operations** on a number of selected cases. They consider proper ones those showing signs of **increased intracranial pressure** as evidenced by papilledema and increased pressure of the cerebrospinal fluid. They opened the skull over the motor area on one or both sides as required. In most of their cases the brain cortex was apparently healthy, meningeal cysts being the lesion found. These were punctured and their walls removed. In 65

cases so treated they report marked improvement in 25, the youngest being two and a half and the oldest seventeen years old. They consider that the earlier the operation is done the better the outlook for improvement. If the *epileptiform convulsions* are *jacksonian* in type, an **exploratory operation** is indicated and cysts or other foci of irritation should be removed. If there is reason to believe that *hereditary syphilis* may be the cause, vigorous **antisyphilitic treatment** should be given.

After *contractures* and *deformities* have developed, varying measures may be employed to lessen their severity. The orthopedic surgeon may do much in proper cases by **breaking up joint adhesions** and by the various **muscle and tendon operations** as required. These should be followed by **passive movements, educational exercises** and **electricity**. When the latter is used it should be applied only to the extensor muscles as the object is to counteract the overacting flexors. In many cases these measures may bring about some improvement without surgical interference. This, of course, will be so only when joint changes and ankylosis have not occurred. **Partial resection of motor nerves** supplying overacting muscles has also been done. When the athetoid spasms are extremely severe, this may give a measure of relief.

In those cases in which the disability is due to spasticity rather than to muscular weakness, Förster's operation of **cutting** some of the **posterior spinal nerve-roots (rhizotomy)** may lessen the disability. Frazier has collected 70 cases of Little's disease in which this was done, with 2 cured, improvement in 56, no improvement in 3, and 9 deaths.

Epileptic convulsions must be treated by the usual methods employed for epilepsy. Mental impairment, if marked, may need custodial care; if of mild degree, something may be done by proper educational measures.

Prognosis.—Stillbirth and death shortly after birth are not infrequently due to meningeal hemorrhage. If the condition is recognized early, there is a possibility of saving life and lessening the future disability by resorting to lumbar puncture. If this produces no improvement, opening the skull by the method of Cushing may be resorted to with some hope of success. A large percentage of those who survive are doomed to more or less disability and a considerable number of these to epilepsy and mental enfeeblement in addition. As regards the former, it should be remembered that it may not appear until the child is several years of age, in fact, may be deferred until puberty. The disability is apt to improve as the child grows older and this may be helped in a degree by employing proper therapeutic measures. The legs as a rule improve more than the arms when these are also affected. It must be remembered that sometimes weakness and spasticity largely disappear while the athetoid or other spasmodic movements remain. Mental impairment, if not of too low a grade, may be improved by proper measures. A certain proportion may become self-supporting, but this is not the rule.

Historical Summary.—Attention was first called to the relation of paralysis and spasticity to abnormal birth in 1861 by Little, the English surgeon. In 1885 Sarah MacNutt showed that meningeal hemorrhage was the lesion in many of these cases. Acute encephalitis as a cause in cases following infectious disease was called attention to by Strümpell in 1884. Batten attributed certain cases of acute ataxia in children to this cause in 1903. The atonic-astasic was described by Förster in 1909 and again by Pierce Clark in 1913, while the ataxic type or diataxia cerebialis infantilis was described by Hunt in 1918. Further historical data can be obtained by reference to the Bibliography.

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(A complete bibliography can be obtained by reference to the books and papers mentioned above, especially those of Osler and Hunt.)

CHAPTER XII

SYPHILIS OF THE NERVOUS SYSTEM

BY JULIUS GRINKER, M.D.

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Introduction.—Among the diseases affecting the nervous system, syphilis occupies an important place. Not only is it frequent, but in its train follow irreparable damage and chronic invalidism. Neurosyphilis—as nervous syphilis is now termed—merits especial consideration because its treatment has become rational and efficacious. And progress is not limited to therapy. It embraces also diagnosis, concerning which it is no longer needful to quote Gowers' statement made as late as 1890: "To the surgeon the processes of syphilis are for the most part open and manifest; to the physician they are secret; its ways are obscure, its language is seldom unequivocal." On the contrary, to-day we are in a position to state definitely that, owing to new clinical and serological methods of diagnosis, syphilis of the viscera, including the

nervous system, has become almost as open and manifest as surgical syphilis.

Neurosyphilis has an extremely varied localization, hence it does not lend itself readily to a description befitting all varieties. Symptoms must differ according to localization of the disease processes within the nerve centers. However, there are certain general features common to all varieties, which may precede the discussion of the separate groups.

Fortunately for our purpose, many of the brain and spinal cord functions have been ascertained, thus warranting definite statements as to the probable seat of a given lesion. Though the task has thereby been made easier, there are still numerous difficulties besetting the path of the diagnostician, for the disease neither limits itself to one location, nor does it always attack one histological structure. Indeed, neurosyphilis is known by its proclivity to attack simultaneously or in rapid succession more than one territory and more than one tissue. It is a study of the grouping of symptoms peculiar to syphilis of the nervous system which frequently leads to the diagnosis when all else fails.

Etiology.—In former times much space was expended in discussing the various theories of the causation of syphilis. Numerous plausible theories have been advanced, which seemed to explain some of the phenomena of this multiform disease. And because of its multiformity, syphilis was held by clinicians and pathologists to be of parasitic origin. The proof for this assumption was the discovery of the *Spirochæta pallida*, an organism also known as *Treponema pallidum*.

In May, 1905, Schaudinn and Hoffmann announced their great discovery of the real cause of syphilis. According to Frederick G. Harris, the organism, as described by Schaudinn, is an extremely slender thread, corkscrew-like in shape and about one millimeter in length, provided with flagella, which stains with most of the aniline dyes. For lack of space we must abstain from a description of the cultural and growth characteristics of the spirochetes, but we may now make the definite statement that the spirochetes are the direct cause of *all* the lesions, not only of some, described as syphilitic. Spirochetes have been demonstrated in all chancres and syphilitic glands, condylomata, mucous patches, and in the blood stream, as well as in syphilids, gumma, and the interstitial connective tissue of the various organs. The organism is found in the great majority of cases of congenital syphilis, not only in the liver—an extract of which is used as the amboceptor in the Wassermann test—but also in most of the patient's organs. And not only is the organism seen in large numbers in the early and secondary lesions, but also in the later stages, the so-called tertiary form, in the heart and blood-vessels, especially the aorta. In 1913, Noguchi and Moore found the spirochetes, or treponema, in brains of paretics and in the cords of tabetics.

Since the causal factors have been found in the pathological tissues of the nervous system, we may well assume that these changes are due directly to the *Spirochæta pallida*.

Why do certain individuals—about 5 per cent. of all those infected

with syphilis—develop neurosyphilis? No one knows. One of several reasons may explain. (1) Special strains of the spirochetes may have a predilection for the nervous system, as shown in those who develop neurosyphilis from a common source. (2) The individual himself may show a special neurotic predisposition, the spirochetes acting as the exciting essential cause. (3) Trauma, excessive wear and tear of the nervous system, sexual and alcoholic excesses may produce a *locus minoris resistentiae* for the spirochetes to invade.

Laboratory Findings.—The most important single diagnostic laboratory test of neurosyphilis is the Wassermann test on blood and spinal fluid. The writer does not purpose to describe the technic of this test, which after all must be left to the trained laboratory worker. It is quite necessary, however, to point out the relative values of positive and negative findings. Often the physician has relied too much upon the opinion of a laboratory technician, when, as a clinician, he is entitled to an independent opinion.

1. **BLOOD WASSERMANN.**—A positive finding in the blood is of great value in the diagnosis and treatment of neurosyphilis, while a negative finding is of little or no value. Fortunately for the reliability and uniformity of this test, the numerous modifications introduced at one time or another have been discarded in most laboratories—nearly all scientific workers have returned to the original test as described by Wassermann.

In early active neurosyphilis 100 per cent. positive Wassermann reactions are expected. In the later stages of neurosyphilis the percentage of positive findings may fall to as low as 65-70 per cent., while in latent syphilis the percentage of positives is still lower—50 per cent. or less. Though there is some doubt that improvement in the blood Wassermann necessarily means clinical improvement, there is no doubt that in the majority of cases clinical improvement carries with it an improvement in the blood-picture.

As a gauge of the *efficacy of treatment* more value can be attributed to spinal fluid findings. It is partly for this reason that lumbar punctures are now being made as a routine accompaniment of the systematic treatment of neurosyphilis.

2. **CEREBROSPINAL FLUID.**—As a means of diagnosis the positive finding of a Wassermann in the spinal fluid ranks high, though not every such finding means that the lesion from which the patient suffers is necessarily caused by syphilis. A patient suffering from syphilis does not thereby become immune from other diseases; but there is a strong presumption that nervous symptoms, appearing in one whose spinal fluid is positive, are caused by syphilis, unless another cause is found. The work of Hauptmann and Nonne has made the spinal fluid examination as a diagnostic measure even more valuable than hitherto. They found that many more positive reactions are obtained when the spinal fluid is gradually increased from the usual amount of 0.2 cu.mm. to 0.4, 0.6, 0.8, up to 1 cu.mm. By this method one finds that in general paresis the reaction is uniformly positive with the small quantities of spinal

fluid—0.2 cu.mm.—while with larger quantities we also get positive reactions in cerebrospinal syphilis and in tabes. Their observations have been confirmed by others and are now utilized for differential diagnosis between general paresis and cerebrospinal syphilis of the interstitial variety, as well as for differentiation between simple tabes and taboparesis, in the latter of which the smallest quantities will produce strong reactions.

It may be put down as a general rule with but few exceptions that in the absence of nervous syphilis, or when such symptoms have disappeared, the cerebrospinal fluid does not yield positive findings. Indeed, the positive findings of Wassermann in the spinal fluid constitutes a true sign of neurosyphilis.

Lumbar Puncture.—First described by Quinke, in 1891, for the purpose of treating the syndrome known as meningitis serosa interna—otherwise known as internal hydrocephalus—lumbar puncture has come to occupy an important position among the necessary steps in the diagnosis of neurosyphilis. The technic for its performance is comparatively simple and practically as first described by Quinke. It should be done with care. The fluid being withdrawn slowly, the patient should remain in bed several hours after puncture, preferably with the head lowered and the foot of the bed raised. These precautions are taken with a view to preventing a disturbance of the brain from the sudden withdrawal of the fluid. One of the disagreeable after-effects following lumbar puncture is *severe headache*, which may last from one to three weeks. There are some individuals who react to lumbar puncture by the development of nausea, vomiting and dizziness; but these persons react unfavorably to almost anything else. An attempt has recently been made to explain the unpleasant effects of lumbar puncture, and a remedy has been suggested. It has been stated that in those cases there is a slow leakage of the spinal fluid into the tissues which drains the entire canalicular system of its contents; the puncture has remained patent, owing to the use of the ordinary thick-calibered lumbar puncture needle. To avoid a constant drain of fluid the use of finer needles has been recommended, a procedure which the writer is now following. For the relief of headache produced by the puncture, 10 grains (0.65 gram) of aspirin, antipyrin, or pyramidon may be administered—remedies which are occasionally useful. In the majority of instances nothing but time brings relief.

Lumbar puncture must not be undertaken unless there is a positive indication for its performance. There are certain real dangers connected with its indiscriminate use. Sudden deaths have been reported after puncture when the patient had signs of brain tumor situated in the posterior fossa. In brain tumor, therefore, regardless of whether it is syphilitic or non-syphilitic, lumbar puncture is contra-indicated. Because of the possibility of an obstruction being present in the communication between the spinal space and the ventricular system, a vacuum may be created in the lower space. The pons-medulla may then be drawn into the foramen magnum, with instant death as the inevitable

result. In one of the writer's cases of definite tumor in the posterior fossa with an urgent need of spinal puncture for therapeutic purposes, the difficulty was overcome by placing the patient in the Trendelenburg position, which he retained during the twenty-four hours following. The fluid was withdrawn very slowly and the patient received the expected benefit—namely, perfect freedom from intense headaches, the result of an over-filled ventricular system.

The spinal fluid should be examined immediately upon withdrawal, as after long standing degenerative changes take place in the cells.

Lymphocytosis.—Normally from 7 to 10 lymphocytes may be found in each cu.mm. of spinal fluid. Whenever the number increases to 15 or more lymphocytes it is considered pathological and is suggestive of the chronic inflammatory changes found in syphilis of the meninges.

Because of the ease with which a cell-count can be made, the Fuchs-Rosenthal chamber has become popular, but an ordinary white blood-cell pipet and the usual counting chamber may be utilized for this purpose.

Normal cerebrospinal fluid is clear, with a specific gravity of 1.006, and possesses a slightly alkaline reaction. It contains but few cells of any kind, the average, according to the Fuchs-Rosenthal method, being from 1 to 5 lymphocytes per cu.mm.

In cerebrospinal syphilis, tabes, and general paresis, the lymphocytes are increased in number. This increase is variable and may run into the hundreds in acute and fulminant cases. Most meningeal inflammations are accompanied by an increase in cellular content, both polynuclear leukocytes and lymphocytes. Lymphocytosis is the expression of a more or less chronic inflammation, while acute forms of meningitis—with the exception of the tuberculous form—are characterized by an increase in the polynuclear cells. The marked lymphocytosis in syphilis is attributed to an extremely chronic type of meningitis.

It has been shown by Siccard, Ravaut, and others, that pleocytosis (another word for lymphocytosis) of marked grade may antedate all other neurologic symptoms by many years. Indeed, lymphocytosis may appear in the cerebrospinal fluid as early as two months after the original infection. Its presence invariably means existing or coming neurosyphilis.

Active therapy has a tendency to influence lymphocytosis; at first the cells become fewer, later they disappear entirely. Patients whose cell-count is not modified by active antisiphilitic treatment are considered candidates for tabes and general paresis. On the other hand, it is understood that the persistent absence of lymphocytosis in the spinal fluid may be taken to mean that the diagnosis of tabes or general paresis is highly improbable. The mere finding of lymphocytosis is not a sign of syphilis, for other conditions may produce lymphocytosis, combined with polynuclear leukocytosis; it may be present in tuberculosis of the nervous system, also in acute anterior poliomyelitis and in herpes zoster.

Lymphocytosis alone is of no pathognomonic value; it becomes important only when associated with other symptoms of nervous syphilis. While treating a patient, the degree of lymphocytosis present constitutes a fairly accurate guide as to the severity of the process. The more effective the treatment of cerebrospinal syphilis, the greater will be the reduction of the number of lymphocytes in the spinal fluid. The same rule does not hold good for the so-called parenchymatous diseases, tabes and general paresis; in the latter there may be a diminution in lymphocytosis, but no corresponding improvement in the clinical picture.

Chemical Examination.—For practical clinical diagnosis and prognosis, the examination of the proteid contents of the spinal fluid is important.

Determination of Globulin.—In cerebrospinal syphilis and in tabes as well as in paresis, an increase of globulin is found in the spinal fluid. As a rule the increase in globulin runs parallel with the lymphocytosis and is independent of the Wassermann reaction. An increase of globulin may be observed in tuberculous meningitis, influenza meningitis, pneumonia, and the several pathological conditions causing irritation of the meninges. Indeed, anything which causes compression of the cord, such as tumor and meningeal infiltration, may also produce an increase of globulin. The importance of an increase of globulin consists in its frequent association with lymphocytosis; only then has it diagnostic value for syphilis.

The methods for the determination of globulin in the spinal fluid are (a) Nonne's test and its modifications, (b) the Ross-Jones method, (c) the method of Noguchi, and (d) Pandey's method.

(a) *Nonne's Test.*—In a small test-tube place 1 c.c. each of cerebrospinal fluid and of saturated solution of ammonium sulphate. Mix well and stand aside for 3 minutes. A normal fluid remains clear or shows but a very faint opalescence; an increase of protein is indicated by opalescence, which may be slight or marked; turbidity or precipitation demonstrates a strong globulin reaction.

(b) *Ross-Jones Test.*—Pour 2 c.c. of saturated solution of ammonium sulphate into a small test-tube. Gently place a layer of 1 c.c. of spinal fluid over the ammonium sulphate solution, the same as for Heller's albumin test. When a white ring is obtained after three minutes, the reaction is positive for globulin excess.

(c) *Noguchi Test.*—Place 0.2 c.c. of cerebrospinal fluid into a small test-tube and add 1 c.c. of a 10 per cent. solution of butyric acid in normal salt solution. Heat to boiling. Add 0.2 c.c. of a normal salt solution of sodium hydroxid and boil once more for a few seconds. The normal reaction is clear or shows a faint opalescence which does not flake for several hours; a positive reaction is indicated by marked opalescence, turbidity, or visible granular precipitate, which settles in about an hour.

(d) *Pandey's Test.*—To 1 c.c. of a saturated watery solution of carbolic acid (1 part carbolic acid crystals and 15 parts distilled water)

in a small test-tube is added 1 drop of clear cerebrospinal fluid. The immediate formation of a bluish-white ring or cloud is the evidence of an abnormal protein content; a white precipitate indicates a strong reaction.

Having briefly described the *four reactions*, namely, Wassermann reaction on blood and spinal fluid, lymphocytosis, and globulin reaction, it is now necessary to discuss their diagnostic values. These reactions, taken singly, mean nothing positive as regards the differentiation of the various types of neurosyphilis, but when read together they offer important guides in diagnosis and treatment.

We shall now reproduce *Nonne's conclusions*, based on large material in the Eppendorf Hospital of Hamburg:

I. *Blood Examination.*

Wassermann reaction:

1. Positive. Is characteristic of syphilis with few exceptions, and indicates that the individual has in some manner acquired the disease, either through heredity or by infection. The disease from which he suffers at present is not necessarily due to syphilis.
2. Negative. Speaks against general paresis, since the disease rarely gives a negative blood-reaction.

II. *Cerebrospinal Fluid.*

1. Normal Fluid. Pressure 90 to 130 cu.mm. water. Globulin reaction negative. No pleocytosis—not over 5 to 6 cells to the c.c. of fluid (Fuchs-Rosenthal).
2. Pathological Fluids.
 - (a) Increased pressure—over 150 cu.mm. water.
 - (b) Globulin reaction positive.
 - (c) Increased cell-count; indicates the presence of an organic nervous disorder, not necessarily syphilitic.
 - (d) If the disease is syphilis, the Wassermann test will be positive on the spinal fluid. If only 0.2 c.c. of fluid are required to give a positive reaction, there is great probability that the patient will suffer from either paresis or taboparesis; it is much less probable that he is the subject of cerebrospinal syphilis or beginning tabes.

In nearly all cases of cerebrospinal syphilis and of tabes the Wassermann becomes positive by the use of larger quantities of fluid—from 0.4 to 1 c.c.

Nonne has also formulated typical findings for the three leading types of neurosyphilis as follows:

I. *Paresis or Taboparesis.*

1. Wassermann reaction on blood positive (100 per cent.).
Pressure increased.
2. Globulin reaction positive (95-100 per cent.).
3. Lymphocytosis (95 per cent.).
4. Wassermann in fluid:
 - (a) Positive in about 85-90 per cent. with original method and 0.2 c.c. fluid.
 - (b) Positive in 100 per cent. with larger quantities of fluid.

II. *Tabes without Paresis.*

1. Wassermann reaction on blood positive (60-70 per cent.).
Pressure usually increased.
2. Globulin reaction positive (90 per cent.).
3. Lymphocytosis positive (90 per cent.).
4. Wassermann in fluid:
 - (a) Positive in about 5-10 per cent. with original method and 0.2 c.c. fluid.
 - (b) Positive in 100 per cent. with larger quantities of fluid.

III. *Cerebrospinal Syphilis.*

1. Wassermann reaction on blood positive (80-90 per cent.).
Pressure frequently increased.
2. Globulin reaction usually positive, exceptionally negative.
3. Lymphocytosis nearly always positive.
4. Wassermann in fluid:
 - (a) Positive in about 10 per cent. with original method and 0.2 c.c. fluid.
 - (b) Nearly always positive with larger quantities of fluid. Of great value in differential diagnosis from multiple sclerosis, cerebral and spinal tumor, which are always negative.

An additional test has been described by Noguchi, the so-called "luetin test," which may become of value in the latent cases of neurosyphilis, that is, those without symptoms and even without other laboratory findings.

The Luetin Test.—This consists of an intradermal injection of killed pallida cultures, as prepared by Noguchi, and called luetin. When the reaction is positive, there will appear, within five or six days at most, a red papule with an indurated areola. The reports on the value of this test are rather contradictory, and not much has been heard from it of late. It is mentioned here for the sake of completeness.

Diagnosis.—As a rule the diagnosis of neurosyphilis is not difficult. This is especially true since we are in possession of the various laboratory tests discussed in the preceding pages. Unfortunately laboratory

workers furnish contradictory reports on cases which require the greatest amount of illumination. It often happens that the diagnosis, previously doubtful, becomes more so after the laboratory has reported. Physicians rely too much upon the laboratory and too little upon their own diagnostic skill. The writer does not deny laboratories the right to assist and corroborate in diagnosis, but the function of the physician is to study every angle of his case and then to do his own thinking. Accumulated clinical experience has taught us that neurosyphilis may mimic almost every organic disease, but we have also learned to detect the actor. It is in the peculiar grouping of symptoms and syndromes that we must seek help in doubtful cases. For instance, a case of thrombotic hemiplegia occurring in a young individual speaks its own language. Thrombosis of a cerebral artery is the special prerogative of the old, unless the young have bought this privilege at the shrine of Venus. Another peculiarity of neurosyphilis is its frivolity: the symptoms have a tendency to change from moment to moment and they are fond of a step-like course. Other organic diseases are more stable and more continuously progressive; syphilis may be known by its very instability. The most serious phenomenon may disappear within a few hours and return the next day or next year. A patient struck with aphasia may recover speech within an hour and may permanently lose it again the same day. Another quality of the syphilitic symptom is its tendency to disregard all classical and textbook behavior; there is something unfinished and incomplete about it. Neurosyphilis acts in defiance of all the rules laid down for the proper conduct of orthodox brain or spinal cord lesions; their very grotesqueness mark them as separate diseases. Regarding the conformity of syphilis to a type, it has been well said that the only thing in which syphilis is typical is that it is *atypical*.

Perhaps the most important clinical guide in the diagnosis of nervous syphilis is the *pupil*. It has been the writer's sign-post for many years, and it has rarely failed to give proper directions. Its language is direct, and when it presents inequalities, irregularities, complete or incomplete Argyll Robertson qualities, it speaks in unequivocal terms. Strange as it may appear, with many the examination of the pupil is still an unknown art. Inequality, loss or exaggeration of tendon reflexes, offer valuable hints to those in quest of assistance and who understand the reading of symptoms.

For purposes of differentiation, before the Wassermann test prevailed in syphilis-diagnosis, writers were in the habit of placing the various symptoms of disease in juxtaposition. All this is unnecessary, since Nonne, a clever observer, has discovered that some of his most classical cases of multiple sclerosis have proved to be cerebrospinal syphilis by the Wassermann on the spinal fluid. The writer, with this knowledge in his possession, picked two cases for clinical demonstration—one a case of multiple sclerosis, the other, a case of cerebrospinal syphilis. The last case showed nystagmus, scanning speech, intention tremor, bilateral Babinski, absent abdominal reflexes, and yet it was proved to be syphilis by the laboratory as well as by the course of the disease towards im-

provement. The first case, with fewer symptoms of multiple sclerosis, was a real case of that incurable disease. This proves the futility of taking up a number of symptoms and comparing them with each other for diagnostic purposes. The diagnosis must be made after a full consideration of symptomatology, course of the disease, and laboratory findings. By laboratory findings is not meant the examination of a specimen of blood only; the writer would especially include the spinal fluid. In neurosyphilis the blood is often negative when the spinal fluid is positive. Much help may be derived from a knowledge of laboratory values as interpreted by Nonne and reproduced in the preceding section. Nonne's "four reactions" have certainly made diagnosis more scientific and have relegated the tedious "therapeutic test" into the past.

Treatment.—GENERAL PROPHYLAXIS.—The general prophylaxis of nervous syphilis is identical with that of syphilis as a disease. Until recently all efforts at prevention consisted in warning the young of the dangers of sexual intercourse and in the regulation and control of prostitution by the police. Such efforts, made rather feebly, have been sterile of results. The public at large has always turned a deaf ear to all serious efforts at the prevention of the venereal peril. It is only from the army and navy that we have received real encouragement in prophylaxis. The percentage of the prevalence of syphilis has been gradually reduced in most armies from 11 to 15 per thousand to 3 or 4 per thousand—certainly a great improvement. The means employed in the army may be classified as educational, legislative and medical. The men are given lectures and literature dealing with the dangers from venereal infection. Having developed a disease, the soldier is required to report immediately; neglect of this order is followed by punishment. In addition, soldiers returning from leave of absence are given preventive treatment by means of the 3 per cent. calomel ointment. For this purpose and for the specific treatment of venereal diseases good hospital facilities have been provided in the army. In this respect the civilian population has fallen behind the military. The real solution of this vexed problem must begin as educational propaganda, not among those unable to benefit by it—namely, old men and women—but among the young. University students, including those studying medicine, should receive **special lectures** on the subject of prevention. **Serological diagnosis of syphilis** should constitute an obligatory study of the required laboratory course. It is almost unnecessary to add that the subject of treatment and prevention of venereal diseases should form a necessary part of all **post-graduate instruction** given in the various postgraduate schools.

In the writer's opinion health and money could be saved the community if the medical examiners of insurance and fraternal orders were instructed to examine as diligently for venereal disease as they do for cardiovascular changes. Not infrequently has the writer been consulted for frank tabes and cerebrospinal syphilis by men who had recently passed rigid examinations for life insurance and were given policies for large amounts. This is a travesty on medicine as a science and an art. Had the insurance examiner taken the pains to tap the knee rather

than feel the pulse, he might have saved his company money and the patient his health.

It has been suggested that something might be gained for prophylaxis if a law were passed making it a misdemeanor to transmit venereal disease to another person. While evidence of such a crime might be difficult to obtain, yet the fear of detection might act as a preventive.

The **public should be educated** as to the gravity of syphilitic infection; those especially who are capable of spreading the disorder by external means must be urged to adopt preventive measures. Midwives, nurses, barbers, waiters, cooks, should be taught how to **sterilize tools and instruments**.

Compulsory reporting of venereal diseases, adopted by most State boards of health, will help not only the individual, but also the community. We shall gradually accumulate a mass of statistics proving the prevalence of venereal disorders and pointing the way toward prevention.

INDIVIDUAL PROPHYLAXIS.—Patients who have already acquired syphilis may do much toward preventing involvement of the nervous system. Scrupulous attention must be paid to the accepted rules of physical and mental hygiene. Not alone is **physical over-work** detrimental, but **mental fatigue and worry** have a tendency to prepare the nervous system for neurosyphilis, the worst form of that dreaded disorder. In addition, **trauma** is to be avoided, because of its peculiar effects on the nervous system in one who is the subject of either syphilis or some other infection. We are all familiar with the clinical observation that trauma often ushers in tuberculosis in the traumatized parts, but it is not so well known that this also applies to syphilis. Localized, luetic, cerebral meningitis has frequently followed an insignificant trauma to the head.

As a matter of individual prophylaxis of neurosyphilis must be mentioned **frequent examinations of the spinal fluid** in one who has contracted syphilis. It has been found that when syphilis has become constitutional, even in the early stages, there is apt to be involvement of the meninges and of the interstitial tissues of the central nervous system. If the condition is detected early, it may be possible, by judicious treatment, to prevent the development of cerebrospinal syphilis, tabes, and general paresis. In order, therefore, to prevent the most serious form of syphilis—namely, neurosyphilis—from appearing many years after the patient has considered himself well, the watchful physician must not fail to urge the **methodical examination of blood and spinal fluid at a time when the patient is free from nervous involvement**.

As the result of such examinations, Corbus, in 1910, found involvement of the cerebrospinal system without clinical symptoms in 33 $\frac{1}{3}$ per cent. of secondary syphilis, 40 per cent. of tertiary syphilis, and 31 per cent. of latent syphilis. In summing up his cases—180 in all—there were 33 (or 30 per cent.) which showed early involvement of the nervous system, though there were no definite clinical symptoms. It seems logical to assume that if these patients are permitted to go untreated, they will eventually show signs of neurosyphilis.

Not only Corbus, but also Fordyce had similar experiences, and the latter therefore pleads with dermatologists and genito-urinary surgeons to pay closer attention to the **early manifestations of nerve-involvement**, as shown by the spinal fluid Wassermann. According to the views of many authorities on syphilis, a **lumbar puncture** should be made when the patient presents himself for the first examination.

In the writer's opinion cases of secondary, tertiary, and latent syphilis should receive **intensive treatment with mercury and salvarsan** even in the absence of all clinical symptoms, provided the spinal fluid is pathological—this as a mere prophylactic measure.

GENERAL MANAGEMENT.—Once the diagnosis of neurosyphilis has been made, there should be **no delay** in instituting treatment. Procrastination here may spell irreparable damage to important nerve tissues.

The treatment of syphilis means more than the relief of symptoms; it includes the judicious management of the most treacherous disease known to the profession: The modern physician cannot afford to harbor the delusion that a stereotyped formula will serve in all cases. Some patients are cured spontaneously, that is, without any treatment; others require but little treatment; while in a large number of cases even the most intensive treatment may be powerless to prevent the oncoming parenchymatous diseases—tabes and general paresis.

To the casual reader a special section on the treatment of neurosyphilis might seem superfluous, for syphilis to him is the same no matter where found. This is a mistake; syphilis is not the same everywhere. Not only is the treatment of nervous syphilis different from that of syphilis in general, but even within the nervous system itself there are regional peculiarities which demand different methods of treatment. The practical guide in the management of a given case of syphilis of the nervous system is the new classification of neurosyphilis into (1) *interstitial* and (2) *parenchymatous types*, each to be discussed in this article under its own heading. (See pages 129 and 148.) The interstitial group includes the lesions formerly classed under cerebral, spinal and cerebrospinal syphilis, while the principal representatives of the parenchymatous group are tabes and general paresis.

It may now be stated definitely that in the treatment of neurosyphilis the most favorable results are obtained so long as the case remains one of interstitial syphilis. The improvement in this group is easily explained by the local peculiarities of the lesions which consist, for the most part, of edema or pressure upon nerve centers—not of actual destruction. Possibilities of a cure exist only when the nerve fibers and nerve centers have escaped actual strangulation. It is for this reason that parenchymatous neurosyphilis, with its great destruction of nerve tissue, offers so few possibilities of recovery. According to some authors, the favorable results obtained in the last group of cases were entirely due to the fact that by the side of definite parenchymatous lesions there were still present remnants of interstitial pathology—inflammations and exudates—which yielded to treatment.

SYMPTOMATIC TREATMENT.—This consists largely in the **alleviation**

of pain and discomfort while waiting for the beneficial effects of specific medication. For instance, a patient who is given treatment for syphilis, but meanwhile suffers from intense cephalalgia and insomnia, cannot wait for the effects of salvarsan and mercury; he is entitled to temporary symptomatic relief by means of **anodynes and hypnotics** judiciously administered.

SPECIFIC TREATMENT.—The drugs used in nervous syphilis are the ones utilized in general syphilis with certain modifications in their application—**mercury, salvarsan** and its equivalents (neosalvarsan, arsenobenzol, novarsenobenzol, arsphenamin) and the **iodids**.

1. **Mercury.**—This classical remedy enters into the treatment of almost every form of syphilis, regardless of what other drugs are being used. The administration of mercury in the form of pills or in solution has been abandoned in favor of modes of administration less likely to produce salivation and gastro-intestinal disturbance.

The form of **mercurial medication** which enjoys great popularity with the profession at the present time is the *inunction method*. This consists in rubbing into the patient's skin a varying amount of mercurial ointment, an average dose being one dram (4 grams) daily. This dose is placed in waxed paper, the full contents of which is to be rubbed by the patient into his skin before retiring for the night, a different part of the body being selected for each treatment. After fifteen to twenty minutes of hard rubbing the greater portion of the ointment is usually absorbed. To allow complete absorption to take place during the night, a flannel bandage is applied to the part and left over night. The parts of the body commonly selected are the flexor surfaces in the following rotation: groins, bend of elbows, popliteal spaces, and the inner surface of the thighs. In order to make the skin more supple, so as to absorb a maximum amount of mercury, a **lukewarm bath** may be taken before each rubbing. Certainly a **hot bath** is necessary every fourth night. Thirty of these rubbings constitute a course of mercury treatment. Between the courses of mercury treatment **iodid of sodium** may be administered in doses beginning with thirty drops of the saturated solution, gradually increasing to 1 dram (4 grams) three times daily after meals. The iodids may be taken in large quantities of water, milk, or carbonated water, and continued for a period of about four weeks. Then mercury is resumed and another course consisting of thirty rubbings administered. Alternating courses of mercury and iodids are to be given during the entire period of active treatment. An old and tried method of treating nervous syphilis is by the so-called "mixed" treatment, which means the simultaneous administration of mercury and iodids during a period of six weeks, which constitutes a course. This is followed by six weeks of complete rest from all active medication. This period of rest is to prepare the patient for another course of treatment by giving him a generous and unrestricted diet, tonics, proper exercise and rest. The next active course of treatment is again followed by a similar period of comparative rest, and this in turn by another period of rest. Two years of such active treatment has been considered adequate, provided blood and spinal fluid Wassermann tests have been made and repeatedly found negative.

A more cleanly preparation than ordinary mercurial ointment is *oleate of mercury*, equally as effective. One dram (4 grams) of a 10 per cent. oleate of mercury is to be used night and morning for four days.

For continuous use the dose is reduced to 1 dram (4 grams) daily, as soon as evidence of salivation has appeared. The oleate is rubbed into the skin by means of a piece of flannel, selecting a different part of the body for each application, but making use of the same piece of flannel. The oleate has one advantage over the mercurial ointment in the fact that it can be used over any part of the body, and another, because absorption takes place more readily. The usual course with oleate is the same as with mercury—six weeks. Rubbings may be ordered alone or in combination with the internal administration of the iodids.

Mercury Injections.—This is a more energetic mode of attacking the disease and appears to be especially indicated when the symptoms threaten to destroy the patient or to cause permanent damage. Depending upon the time at our disposal, we make use of either soluble mercurial salts frequently repeated, or of the insoluble mercurial preparations repeated at longer intervals.

Soluble Mercurial Salts.—The soluble mercurial salts, such as mercuric chlorid, succinamid and the red iodid of mercury, are injected daily, or every other day, in doses varying from $\frac{1}{8}$ of a grain to $\frac{1}{2}$ grain (0.008-0.0324 gram). In fulminant cases of brain and spinal cord syphilis, which suggest impending or already existing apoplexy, in stupor or coma of syphilitic origin, the soluble and quickly acting mercurial salts are indicated. All antiseptic precautions must be observed, needles and syringe must be boiled and the physician's hands made sterile. Fresh tincture of iodine applied to the skin and followed by alcohol is usually sufficient to sterilize the skin over the buttocks. The injection is made directly into the buttocks, and one should aim to avoid piercing blood-vessels and nerves. This can be accomplished by choosing as the site of intramuscular injection a point halfway between the anterior superior spine of the ilium and the upper end of the infragluteal fold. This point brings one well outside the reach of important blood-vessels and nerves.

Insoluble Mercury Salts.—When time is not an element of great importance, as in the chronic forms of parenchymatous neurosyphilis, the insoluble salts of mercury may advantageously find application. The most popular representatives of that class are salicylate of mercury and the so-called "gray oil," each having advocates. The writer prefers the gray oil prepared by the National Pathological Laboratory and sold in syringes containing sufficient for ten injections. This preparation is painless and easily administered. The dose of both drugs is approximately one grain (0.06 gram), once or twice weekly, injected into the buttocks. The landmarks are the same as those previously given for the injection of the soluble mercurials.

The advantage of the insoluble preparations over the soluble ones lies in the slowness of absorption—the system being fed with small quantities of mercury during the intervals between injections—while the soluble salts are rapidly excreted and require frequent repetition. One disadvantage in the use of the insoluble preparations is the absence of control over the rate of absorption, which must vary in different individuals. A knowledge of the uncertainty of the rate of absorption makes imperative frequent examinations for signs of beginning salivation. On the appearance of reddened or spongy gums, or of a mercurial breath, injections should be discontinued. Indeed, it is a good rule to interrupt the treatment after each series of eight injections, the ob-

ject being to await the possible development of signs of mercurialization.

The patient must be impressed with the fact that, irrespective of the mercurial preparation used, he must give careful attention to the oral cavity: teeth and gums require brushing after each meal. Powdered chlorate of potash may be used as a tooth-powder, and listerine or a similar antiseptic as a mouth-wash. During treatment with mercury all articles containing even a trace of organic or mineral acids should be excluded from the dietary, which means that raw or cooked fruit is also to be prohibited. These precautions are to be followed to the letter, as otherwise there will be salivation with all its discomforts, which may necessitate cessation of much needed treatment for a long time.

2. *Tartrobismuthate of Sodium*.—In 1922 Marie and Fourcade reported on the treatment of neurosyphilis by means of tartrobismuthate of sodium. Their method consisted in the administration of one-and-one-half c.c. of the salt intramuscularly every five days, until 20 injections have been given, which approximately represents three grains of the salt for each patient.

In their cases the results were absolutely *nil* for the fully-developed cases of general paresis, while there was some improvement in the early simple forms of general paresis. The drug appears to exert a favorable influence on the gummatous and vascular types of neurosyphilis, and favorable reports have come from a number of reliable observers of different countries. "Watchful waiting" is a good motto to follow with reference to all the bismuth preparations.

3. *Salvarsan and Neosalvarsan (Arsphenamin and Neoarsphenamin), Arsenobenzol, Novarsenobenzol, Silver-arsphenamin, Sulpharsphenamin, Tryparsamid*.—Preceding a discussion of the uses of these arsenicals in nervous syphilis, it is necessary to give a short sketch of such preparations as have only *recently* come into general use, and of others about to be brought to our notice soon.

(a) *Silver-arsphenamin*.—This is thought a good substitute for arsphenamin and neoarsphenamin in cases with severe reactions from the use of the latter. Some have not only found silver-arsphenamin as effective but even superior to the older arsphenamins. Though its arsenic-content is only two-thirds that of arsphenamin, it is nevertheless two and three times as active because it contains 12.7 per cent. of silver. Like the other preparations, it is sold in ampoules of various sizes. Doses vary from 0.15 to 0.25, or 0.3 gm., usually administered in 5 to 10 c.c. of freshly distilled water. A course of treatment consists of 8 to 15 injections given at intervals of four to five days. Karl Stern's simple technic for intravenous injection of silver-arsphenamin is, to dissolve the black powder in 2 to 3 c.c. of warm water contained in a 10 c.c. syringe. After puncturing the vein, the piston is withdrawn until the syringe is full of blood and remedy before being pressed home. The admixture of albumin which takes place in the syringe is believed to reduce the toxicity of silver-arsphenamin. Of this preparation, concentrated solutions are also well tolerated intramuscularly.

(b) *Sulpharsphenamin*.—This is an arsenobenzol derivative, prepared from arsphenamin, formaldehyd, and sodium bisulphite. It possesses great stability in dry form and in solution. In contrast to arsphenamin and neoarsphenamin, both of which rapidly change in color and composition on standing in solution, sulpharsphenamin remains clear and shows no increase in toxicity after twenty-four hours' standing.

The average arsenic-content is 22 per cent., while that of arsphenamin is 30 per cent. Sulpharsphenamin has not only been found to be more stable than other arsphenamin preparations, but it also possesses greater tissue penetration. Biologically it displayed practically the same percentage efficiency in its effect upon trypanosomes in the spinal fluid as tryparsamid (tryparsamid 87 per cent., sulpharsphenamin 82 per cent.). Sulpharsphenamin may be injected intravenously and intramuscularly with equally good results. Ampoules consist of various-sized doses, from 0.3 to 0.6 gm., containing a readily soluble, light-yellowish powder. The dose is gradually increased from 0.3 to 0.6 gm. which is considered the maximum dose. Intravenous injections are given once weekly in more or less concentrated solution, depending on the operator's inclination. For intramuscular injections, highly concentrated solutions are best tolerated. For this purpose a 30 per cent. concentration is recommended, that is, for each 0.1 gm. sulpharsphenamin 0.3 c.c. of water is required. The intramuscular injection may be given into the gluteal region with a 2 c.c. hypodermic syringe. With the sulpharsphenamin treatment is usually combined the regular mercury salicylate or "gray oil" injection, best given during intervals of treatment with sulpharsphenamin. Dr. R. D. Halloran of the Boston State Hospital has pointed out that some of his patients have shown signs of kidney irritation in the form of traces of albumen and occasional hyaline casts after they have completed a course of sulpharsphenamin treatment consisting of seven or eight injections. This, of course, is good warning and should be heeded by those using sulpharsphenamin; frequent urinary examinations are therefore imperative. Carl Veogtlin has come out strongly in favor of sulpharsphenamin as an important rival of the old arsphenamin group.

(c) *Tryparsamid*.—This new arsenical was developed in the Rockefeller Institute for Medical Research. The dried salt contains 25.32 per cent. of arsenic in the pentavalent form, and is a colorless, odorless powder readily soluble in water. A number of the institutions for mental cases have been given an opportunity for clinical experimentation and several reports have already been published, some favoring the use of this drug, while others are still doubtful. Perhaps the most optimistic report on tryparsamid emanated from the Wisconsin University, and was furnished by Lorenz, Laevenhart, Beckwenn, and Hodges. They published a series of cases of general paresis, 54 in all, claiming brilliant results from the administration of tryparsamid intravenously, combined with mercury intramuscularly, without any intraspinal medication. Their treatment consists in dissolving 3 gm. of tryparsamid in 10 c.c. of freshly distilled water and injecting the total amount intravenously; the solution is injected at intervals of a week for eight successive weeks. At the same time mercury salicylate in one-grain doses is administered by intramuscular injection. A mercury injection is given three days before each dose of tryparsamid, and a total of nine such injections alternating with the eight injections of tryparsamid comprise a course. After resting from 5 to 8 weeks, a second course is given. After the second course and a period of rest, or if the case is still serologically positive, a third course is given. Of their 54 cases of general paresis, 28 were discharged from the hospital and have been able to return to work, for periods varying from 6 months to two years. The U. S. Public Health Service has been able to prove experimentally that tryparsamid has greater penetrating power than arsphenamin and neoarsphenamin and other arseni-

cal preparations. The authors also publish tables to prove that tryparsamid has a marked effect on the blood and serology of general paresis, reducing the strength of the positive Wassermann and in many cases producing negative reactions. In the writer's opinion the pernicious effects of tryparsamid on the optic nerves, causing optic atrophy in a large number of cases, constitute a good reason for the most extreme conservatism in the use of what appears to be an otherwise splendid remedy and good substitute for the Swift-Ellis treatment.

Discussion.—When the arsphenamins were first introduced by Ehrlich, an impression prevailed that its spirocheticidal powers were exerted against only the early lesions of syphilis. Most authors, including Ehrlich himself, warned against its use in the late lesions, especially in those affecting the central nervous system. Cautiously he advised the experimental use of small doses of the remedy in all forms of neurosyphilis. Soon a series of phenomena were observed, which were variously described as neurorecidives and neurorecurrences. These sequelæ were characterized by affections of the cranial nerves accompanied in most instances by deafness and cranial nerve paralysis. During the period of the administration of salvarsan in small doses the symptoms described as neurorecurrences multiplied at an enormous rate. It was then suggested that possibly the small doses administered failed to destroy the spirochetes, but caused them to multiply instead. While controversies as to the true nature of these relapses were carried on, some observers reported that the administration of larger doses had the effect of removing the unpleasant complications. Simultaneously with this discovery it was found that salvarsan or neosalvarsan, in combination with mercury, was more effective than was either remedy administered alone. When subsequently the proof was produced that all forms of luetic nervous disease are syphilis—not merely related to it—the hope was cherished by many that henceforth all forms of syphilis would be treated alike. This was found to be deceptive. It is true that certain of the early syphilids yield readily, within a very short time, to one or two intravenous injections of salvarsan, but this is no indication that cerebrospinal syphilis can be cured in the same way. On the contrary, it has been definitely ascertained that only fair-sized doses, frequently administered, exert any appreciable effect on the late lesions of syphilis, notably neurosyphilis. Conformably to this view, Collins and his co-workers advised the so-called "intensive" method of treating nervous syphilis. This method aims to flood the system with salvarsan or neosalvarsan intravenously, allowing two days only between injections, five of which injections are thus administered, unless contra-indications are present. During the intervening days the patient receives inunctions of mercury or intramuscular injections of salicylate of mercury in one-half to two-grain doses (0.0324-0.13 gram) once or twice weekly; or bichlorid of mercury in doses of $\frac{1}{8}$ - $\frac{1}{4}$ grain (0.008-0.016 gram) is injected every day or every other day. After a course of this treatment, extending over three weeks, an examination is made of blood and spinal fluid to decide the further management. If the Wassermann test is negative, treatment is interrupted for three months, at the expiration of which time the series of injections are repeated. This plan of treatment is consistently adhered to until all clinical evidences of syphilis have disappeared and, in addition, the blood and spinal fluid have become negative.

The intensive method of treating neurosyphilis certainly brought

favorable reports, but there were many disappointments. As many relapses seemed to have occurred after this method as after the old line of treatment with mercury only. It was argued that there probably exists an anatomical barrier to the free transmission of salvarsan from the general circulation into the central nervous system, which was made more probable by the fact that experimenters failed to detect any appreciable quantity of salvarsan in the subarachnoid space after large intravenous injections. According to Goldman and others, the choroid plexus, which constitutes the greatest source of the spinal fluid, functionates somewhat like a filter in that certain poisons—salvarsan and neosalvarsan among them—are not permitted to pass into the ventricular system, while the fluid elements are given free passage. Though this peculiar arrangement serves well as a defense against the entrance of poisons into the nervous system, it also prevents the entrance of needed remedies. Marinesco, Robertson, and particularly Swift and Ellis, endeavored to overcome this disadvantage by carrying on similar experiments, in order to discover a method whereby it would be possible to reinject into the subarachnoid space the patient's own serum previously charged with a full dose of either salvarsan or neosalvarsan, or their equivalents. To the ingenuity of Swift and Ellis we owe our present method of intraspinal therapy.

Swift-Ellis Method of Intraspinal Injections.—The patient is given an intravenous injection of 0.6 gram (9 grains) of salvarsan or of 0.9 gram (14 grains) of neosalvarsan in the usual manner. One hour after the injection, enough blood is withdrawn from the patient's vein to yield at least 15 c.c. of serum. The blood, obtained under aseptic precautions, is permitted to coagulate and then placed in an ice-chest over night. Next morning the separated serum is carefully decanted off into a centrifuge tube and allowed to centrifuge for half an hour. The clear supernatant fluid is pipeted off from the few red cells at the bottom and poured into a graduated tube up to the 12 c.c. mark, and then brought up to 30 c.c. by the addition of sterile 0.9 per cent. sodium chlorid solution. This is placed in a 56° C. thermostat for 30 minutes and the mixture of serum and salt is ready for intraspinal injection.

The solution is injected at body temperature, with the patient lying on his side near the edge of the bed. The skin of the back is rendered aseptic, and the area to be punctured is anesthetized with a 2 per cent. sterile novocain solution. A lumbar puncture needle is introduced in the usual manner, and about 30 c.c. of cerebrospinal fluid is withdrawn, or a quantity which will reduce the intraspinal pressure to about 30 to 40 mm. of water. This is gauged with a 3-millimeter glass tube graduated in centimeters and millimeters, which gauge is disconnected when the desired pressure is reached. The use of a gauge is not essential, the only requisite being that the quantity removed equals the quantity introduced. For the past eight years the writer has followed the routine method of removing 5 c.c. less fluid than is to be introduced. The reason for this procedure is to keep the salvarsanized serum slightly under pressure and thus to cause better diffusion throughout the central nervous system. The serum-salt mixture is poured into a Luer syringe (large size), carrying at the delivery point a sterile piece of connecting rubber tubing about twelve inches long. This tubing is then attached to the lumbar puncture needle, taking care not to introduce air. The mixture is then permitted to flow gently into the subdural space. During

the withdrawal of spinal fluid the patient may experience discomfort, in which event a smaller quantity of fluid is withdrawn and the salvarsanized serum injected. Patients must remain in bed twenty-four hours after each intraspinal injection. In order to prevent headaches and disagreeable nausea following injection, the foot of the bed is elevated about six inches and the pillows are removed from under the head. When these precautions are observed, patients usually experience few or no after-effects; perhaps only a slight headache is felt, or there is a slight elevation of temperature within half an hour. In the majority of cases pains in the lower extremities appear within 2-4 hours after an intraspinal injection.

The original technic, as described by the authors, has been somewhat modified of late, especially with reference to the time of blood-withdrawal and as to the dilution with normal salt solution. McCaskey advises shortening the period of the withdrawal of blood to 20 minutes, the object being to increase the salvarsan content. This modification has been generally adopted. Another modification of the Swift-Ellis treatment consists in the use of pure serum, undiluted with normal salt solution, but otherwise according to the authors' directions. In the writer's own practice, 12 c.c. of the undiluted serum, prepared according to Swift-Ellis, are used, which dose is gradually increased with each subsequent injection to 15, 18, 20, 25 and, lastly, to 30 c.c. of undiluted serum.

Direct Intraspinal Injections.—Numerous suggestions at simplification of the Swift-Ellis method have been made by various clinicians. Attempts have been made to introduce salvarsan and neosalvarsan into the subarachnoid space directly without a preceding intravenous injection. Wechselsemann was the first to introduce a small amount of salvarsan intraspinally. Then came Marinesco, Ravaut, Schubert, Genenrich and Wile. It was Wile who popularized Ravaut's method of direct intraspinal medication in America. However, when this short cut to the intradural space had become popular, unfavorable reports began to pour in, due partly to defective technic, but mostly to inherent faults of the method itself. Among the accidents resulting from this method must be mentioned paralysis of legs, bladder and rectal sphincter, as well as decubitus. Corbus, Gordon, Sachs, Strauss and Kaliski have also reported unfavorable results. While the writer has seen brilliant results in one or two hopeless cases, failures were numerous and apparently the result of the direct intraspinal injection of salvarsan and neosalvarsan. The method has been almost entirely abandoned by the medical profession. Most clinicians have returned to the more complicated—but far safer—autoserosalvarsan therapy of Swift-Ellis.

Ogilvie's Method.—Of the numerous contributions to intraspinal therapy, that furnished by Dr. Ogilvie is perhaps as important as the Swift-Ellis method itself. Ogilvie's method consists in the addition of small amounts of salvarsan to human serum previously prepared according to Swift-Ellis, without the intravenous injection of salvarsan. The object of this method is to inject intraspinally a known dose of salvarsan, instead of relying upon an uncertain quantity of the remedy contained in the Swift-Ellis injection. Because of the occasional occurrence of temporary bladder disturbances from larger doses, the author of this method, as well as others who have tried it, warn the profession not to exceed the dose of 1 milligram. It was the large doses that caused

dangerous sequelæ to appear. So favorably has this method been received that Swift himself, in commenting upon Ogilvie's modification, admits its greater spirocheticidal effects as compared with his own method. He believes, however, that a certain as yet unexplained principle derived from the patient's own blood and probably the result of the action of salvarsan upon the blood-constituents—absent in Ogilvie's method but present in his own procedure—is responsible for some of the beneficial effects. Further, in his opinion the administration of *auto-serosalvarsan* to which salvarsan has been added, according to Ogilvie, constitutes probably the ideal method.

Byrnes' Method.—This method is similar to the preceding, differing from it only in the fact that to blood-serum—prepared as for Swift-Ellis without the previous intravenous salvarsan injection—there is added a small dose of mercuric chlorid instead of salvarsan. Byrnes himself believes that the beneficial results reported from the Swift-Ellis treatments are not due to the infinitesimally small amount of salvarsan contained in the 30 c.c. of diluted serum, but rather to the bichlorid of mercury still circulating in the blood and thus transferred directly into the subarachnoid space. He believes that, inasmuch as salvarsan therapy is nearly always combined with energetic mercury medication, it is uncertain which of the two drugs exercises the beneficial effects. He therefore advocates the direct introduction of mercury into the subarachnoid spinal space in doses of from 1/50 to 1/20 of a grain (.0013-.003 gram). The writer, having given this method an extensive trial in hospital practice, is fully convinced of its efficacy in improving laboratory and clinical findings, but, like others, the violent reactions have discouraged him from its further use.

Intracranial Injections.—Two methods of bringing the spirocheticidal substance directly into contact with the brain have been introduced into therapeutics—the subdural and the intraventricular routes. Drew M. Wardner first described the intracranial injections of salvarsanized serum, believing that the usual administration of mercury and salvarsan, intravenously and intraspinally, does not reach the ventricular system, and that, therefore, the most logical treatment of spirochetel involvement of the brain must be directed to the brain itself. This method presents difficulties and has resulted in fatalities.

Ayer's Intracistern Route.—The route through the cisterna magna, first described by Ayer in conjunction with Weggeforth and Essick in 1919 and again by himself in 1920, is less dangerous and perhaps quite as efficacious as the intracranial method. According to Ayer, the procedure has been found almost easy, and no alarming symptoms have been reported either during or after the puncture.

"The patient is placed on the side, as if for lumbar puncture, with neck moderately flexed. Care is taken to maintain the alignment of the vertebral column to prevent scoliosis and torsion. After antiseptic preparation of the skin, usually including the shaving of a little hair, and local anesthetization with procain, the thumb of the left hand is placed on the spine of the axis, and the needle inserted in the midline just above the thumb. The needle may be pushed rapidly through the skin, but should then be cautiously and guardedly forced forward and upward in line with the external auditory meatus and glabella, until the dura is pierced. If the cisterna be entered at this angle there is usually a distance of from 2.5 to 3.0 cm. between dura and medulla as shown on

frozen sections; with the needle less oblique in position the distance between the walls of the cisterna becomes progressively less. Therefore, it is good practice to aim a little higher than the auditory meatus, and, if the needle strikes the occiput, to depress just enough to pass the dura at its uppermost attachment to the foramen magnum. At its entrance the same sudden "give" is felt as in lumbar puncture. The needle employed is a regular lumbar puncture needle, nickloid, 18-gauge preferred, with beveled stylet, sharp on the sides, but not too sharply pointed. There is rather less variation in the depth of the tissue traversed than in the lumbar region, being in an ordinary sized adult from 4 to 5 cm., the greatest distance in the series being 6 cm. and the smallest 3.5 cm. It was found that a faint circular scratch on the needle, 6 cm. from the tip, was entirely satisfactory in judging the distance."

In spite of the simplicity of the technic, the author thinks it unfair to the patient to perform cisterna puncture without previous experience at the necropsy table.

Ayer himself and others after him utilized the intra-cistern route for diagnosis at first, but recently a number of favorable reports have been published on the use of this route for the introduction of arsphenaminized serum (Swift-Ellis technic). Thus, F. G. Ebaugh reports on a series of 250 punctures in 28 paretics. He thinks this method is superior to the intracranial method because of the facility with which it can be performed; in addition he emphasizes the fact that treatment is more intensive than by the intraspinal method, there being less dilution and more widespread dissemination of the serum. The serum reaches all parts of the brain, thus exerting a favorable influence on the syphilitic foci, interstitial and parenchymatous.

Spinal Drainage.—Gilpin and Early in 1915 reported favorably on their method of treating neurosyphilis by means of mixed treatment, namely, mercury and intravenous injections of arsphenamin, followed by complete drainage of the spinal fluid. The technic is simple: The intravenous injection of any of the arsphenamin preparations being completed, a spinal puncture is made and fluid withdrawn until no more flows from the cannula. Spinal drainage should not be performed oftener than once in two weeks, though salvarsan injections and mercurial inunctions may be ordered as before. The headaches are best prevented by the recumbent position in bed for at least twenty-four hours, with head low and the feet slightly elevated. This method has been popularized by Dereum and his co-workers and has become known as Dereum's spinal drainage; he believes this to be equal, if not superior, to the Swift-Ellis treatment. This opinion is not shared by the writer of this article, who is more than ever convinced of the efficacy of the Swift-Ellis method, while he has never seen any striking results from spinal drainage, except the immediate disabling effects in the form of severe headaches.

3. *Iodids.*—Formerly great curative powers were attributed to the iodids. And it was the custom in serious cases of brain syphilis to crowd the iodids in enormous doses. Recently we have learned that iodids have a low spirocheticidal value and their use in most forms of nervous syphilis has been dispensed with. Authors like Collins, Weisenburg and Cotton advise against the administration of iodids altogether, while others have become apathetic towards this form of treatment. There still remain those who, like Jelliffe, have had proof of the efficacy

of iodid administration in the past and who are therefore loath to discontinue its use entirely. The writer agrees with Jelliffe and is still in the habit of using iodids in the treatment of nervous syphilis. While admitting their low spirocheticidal power, one must concede the useful quality of iodids in causing the absorption of inflammatory products resulting from microbial activity. Sometimes great benefit is experienced from the administration of fair-sized doses of iodids in syphilis of the nervous system—a group in which the iodids have celebrated their greatest triumphs. The dose may not exceed 1 dram (4 grams), three times daily, well diluted in water or milk, and taken after meals. Those who still use the so-called “mixed” treatment certainly include the iodids, but in order to conserve the patient’s gastric functions, moderate-sized doses should be administered. The writer’s preference is for the 30-grain dose (2 grams) of the sodium preparation, though recently the opinion has been advanced that potassium iodid is the more valuable preparation for late syphilis.

In many cases Sajodin, a tasteless preparation in tablet form of 0.5 gm. each given three times daily, may be substituted for the ill-tasting iodids.

PLAN OF TREATMENT.—There is no plan of treatment suitable for all cases of neurosyphilis. Something must always be left to individual initiative in medicine. Every clinician has his favorite plan. However, all are agreed that it is well to have a **complete record of laboratory data** before beginning treatment: Wassermann tests on blood and spinal fluid, globulin tests and lymphocyte count. This is necessary not only to make the diagnosis more certain and perhaps to disarm possible criticism, but also as a gauge for comparison in the future. In order to learn something about the efficacy of our treatment, it will be necessary during the course of such treatment to make **spinal fluid tests** at stated intervals.

Authorities are now agreed that the selection of salvarsan or of any of its equivalents is a matter of individual preference; it is not the preparation so much as the proper **technic** which counts. The various new salvarsan preparations are of about equal therapeutic value, provided sound judgment is used in their application. The question of the degree of dilution seems to have been settled in favor of the low dilution and the concentrated solution. The writer’s preference has always been for the concentrated solutions, which have never given occasion for regret. How often shall an intravenous salvarsan injection be given? Not oftener than once weekly. What is the dose? The writer has never seen any ill effects from the regular dose of 0.6 gram (9 grains) salvarsan or 0.9 gram (14 grains) neosalvarsan, though smaller doses have been advised for most forms of nervous syphilis.

Is salvarsan to be given alone or in combination with mercury? The combined treatment, salvarsan and mercury, has been silently adopted by all. It is still questionable as to which of the two drugs exercises the greater spirocheticidal effect on syphilis. All of us have seen marvelous results from the energetic administration of mercury before the salvarsan era; then why discard our ally? Our results are certainly better to-day than they were before we knew salvarsan; we therefore give both, and enough of each.

When are the iodids to be administered in neurosyphilis? Though the iodids have fallen from their high pedestal as the great specific for

nervous syphilis, they act very beneficially as the scavengers of the products of syphilitic exudates, and should be given an opportunity in all inflammatory forms of neurosyphilis. This means that most forms of interstitial nervous syphilis may be considerably aided by the administration of iodids in moderate doses, say 30 grains (2 grams) three times daily.

In which cases of neurosyphilis are intraspinal injections of salvarsanized serum indicated? It is not necessary to apply the Swift-Ellis treatment in the strictly cerebral varieties of interstitial syphilis, for we have seen splendid results from the intravenous salvarsan injections, combined with large doses of mercury and moderate doses of iodids. There can be no question, however, that cerebrospinal syphilis of the chronic variety, especially the cases which fail to respond to the intravenous method, should be subjected to the method of Swift-Ellis.

How often may the intravenous-intraspinal injections be repeated? In the author's opinion, one injection every 10 days, and plenty of mercury during the interim, is good treatment. Only in desperate cases, when the entire house threatens to collapse, and every effort to stop the conflagration has failed, are we justified in administering a Swift-Ellis treatment every few days. Reliable observers have reported cases of sphincter and leg paralyses from too frequent intraspinal injections. Besides it is a good rule to give the patient a rest from these treatments after each series of twelve intraspinal injections.

In concluding the section on the general treatment of neurosyphilis, a statement frequently made, but often forgotten, may be reiterated that each case requires individual consideration and all the light that can be thrown on it by our past experience. The time for following stereotyped formulæ has passed not only for medicine, but also for neurology, and more especially for the treatment of syphilis of the nervous system.

Prognosis.—This depends largely upon an early diagnosis and treatment, for the earlier a case is diagnosed and treated, the better the chances are for a cure. In many cases the recovery is such as to enable patients to resume their occupations.

As long as the changes in the tissues are limited to interstitial structures, the prognosis is good. No sooner is the parenchyma itself involved—which means degeneration of nerve structures—than complete recovery becomes impossible. Likewise, the series of secondary changes occurring in syphilitic arteries are apt to become permanent. The prognosis is hopeful in neurosyphilis caused by inflammation or gumma, except perhaps after optic neuritis, or choked disk, when followed by atrophy of the optic nerve with consequent impairment or loss of vision.

In a general way we are justified in the statement that meningitis, cerebral, spinal, or cerebrospinal, is amenable to cure. In that large class of cases accompanied by paralysis, monoplegia, hemiplegia, or paraplegia, the prognosis is doubtful as to complete recovery, for these lesions, if persisting, indicate permanent and incurable disease. Of course, the paralyses of one or more cranial nerves due to meningitis at the base are almost always favorably influenced by treatment. The wonderful effects of antispecific treatment on certain spinal lesions are also accounted for by the ease with which the underlying condition can be removed, namely, the syphilitic meningitis.

The prognosis must be guarded in cases of cerebral syphilis with

mental symptoms, convulsions or epileptiform attacks. Most unfavorable is that variety of neurosyphilis which progresses toward involvement of the cortical parenchyma, namely, *general paresis*. In this disease there is not only destruction of cortical cells and of tangential fibers, but also widespread cerebral meningitis. The prognosis will depend upon the kind and pathology, and whether the inflammatory changes preponderate over the degenerative ones. The writer believes many of the so-called "cured" cases of general paresis have been pseudo-paresis, that is, meningitis of the frontal lobes with little or no pathology in the parenchyma of the brain; there were clinical symptoms resembling general paresis, but not the disease itself. With this fact in mind, one is not justified in abandoning paresis to its fate, but hoping to overcome the inflammatory (interstitial) changes, we resort to active intraspinal or intracranial antisyphilitic treatment.

On the other hand, not all cases of neurosyphilis showing but few symptoms are necessarily improved by treatment. It is known that some advanced cases of brain syphilis manifest themselves by few symptoms, perhaps only by persistent headache, transient aphasia, monoplegia, hemiplegia or convulsions. Of course, treatment is helpful even in these cases, when begun early and carried on systematically.

The current statement, that when symptoms are caused by syphilis the prognosis is always good, is fallacious. To mention only one type of neurosyphilis, namely, *thrombotic softening*—when this has once occurred in nervous tissues, restoration is impossible, the same as when caused by non-specific softening.

It must be repeated that the appearance of ptosis, diplopia, or facial paralysis does not always mean permanency or unfavorable prognosis; it may mean meningitis at the base of the brain, pressure on cranial nerves. But when these cranial nerve palsies are caused by a permanent blocking of the blood supply to the nerves, the prognosis as to functional recovery is unfavorable.

If basal meningitis were always unilateral, or were to appear independently of arterial changes, the prognosis would always be favorable. As these two types of lesions, namely, *meningitis* and *arterial disease*, often appear side by side, the prognosis is doubtful. Improvement may occur in some symptoms, but others may persist indefinitely.

Involvement of the medulla by the syphilitic process usually spells disaster. The so-called *pseudobulbar variety* of neurosyphilis, like the non-specific case, has few possibilities for recovery, caused as it is by two separate attacks of hemiplegia—one on either side.

When *epilepsy* of syphilitic etiology has become thoroughly established, antispecific remedies have little or no effect upon the epilepsy itself, though the convulsions may be influenced in the same manner and by the same remedies as in non-specific epilepsy. It is well, however, to combine with luminal in regular doses the antispecific treatment for syphilis proper, in the hope that possible meningitic infiltrations over cortical cells may thus become absorbed and the epileptiform disease cured.

The prognosis in all forms of syphilis of the nervous system must be given with caution and much reserve, for of all organs affected by this disease the delicate nervous system suffers most. Only when a patient has been free from all clinical symptoms for a period of five years, and when the blood and spinal fluid have been repeatedly nega-

tive, may we consider him cured. The mere negative findings of blood and spinal fluid in themselves do not justify a favorable prognosis, though they constitute a good index for treatment. Too frequently the positive laboratory findings and clinical symptoms have returned in greater force than ever, after negative laboratory findings have been reported.

In some instances, irrespective of the length of time during which a patient was treated, there will be left a degree of mental deficiency or some form of paralysis. Besides, convulsive seizures and mental troubles have appeared even months and years after all treatment had been discontinued and the patient declared cured.

Gummatous tumors, if of considerable size, require, first, surgical treatment, the same as non-specific neoplasms, and afterwards anti-specific treatment. The prognosis will depend upon the accessibility of the growth and the effect of treatment.

If the prognosis is to remain good, the patient who has once acquired neurosyphilis must be under medical care during the rest of his natural life. At stated intervals laboratory and clinical examinations must be made and courses of treatment administered.

Pathology of Syphilis.—In spite of the enormous amount of work done on this subject, the exact pathology of syphilis is still to be written. However, we are beginning to understand something of the pathogenesis of the disease. Most observers now believe that, dependent upon their early localization in the body, spirochetes select their future course of activity in the individual during the so-called secondary period. In this sense syphilis may be considered as a combination of local infections dating back to an early period in its history, rather than as a chronic infection extending over years. A careful study of the disease makes this theory the more plausible, since we know that the nervous system may become involved in the primary and secondary stages, as shown by an examination of the spinal fluid, which often shows marked lymphocytosis, globulin increase, and positive Wassermann reaction. It is generally believed that patients in whom the spirochetes are found in the nervous system at an early stage are more prone to develop syphilis of the nervous system in the later stages. While it must be admitted that in both the primary and secondary stages of syphilis, tests have given positive results which later became negative, the general rule is that spirochetes localize early and produce their lesions *in situ*. From this statement it appears that the evidence of the localizing powers of the spirochetes assumes great importance. There may also be a peculiar tendency for certain varieties of spirochetes to produce certain lesions—for instance, a general syphiloderma is often followed by local lesions in many organs of the body. Others produce their early effects on the nervous system, and henceforth remain here to expend their fury later. If this is admitted, the practical inference is to be drawn that a systematic examination of the spinal fluid is imperative in every case, in order to prevent, if possible, the early localization in the nervous system. A practical point in therapeutics to be deduced therefrom is, that in the event of general measures of treatment failing to effect a cure, local treatment must be substituted, namely, intraspinal injection of mercury or salvarsan, or both.

In concluding this short outline of the pathology of syphilis, it may be well to quote in abstract the leading American worker on the pathological anatomy of syphilis, Warthin, who maintains that gumma is

not the essential typical lesion of old or latent syphilis. He believes gumma is a relatively rare formation and that the great majority of cases of syphilis run their course without the formation of gummatous granulomas. He further maintains that the essential tissue lesion of either late or latent syphilis is an inhibitive or inflammatory process. This is usually mild in degree and characterized by lymphocytic and plasma infiltrations in the stroma, particularly about the blood-vessels and lymphatics, beginning with slight tissue proliferation, eventually fibrosis and atrophy or degeneration of the parenchyma. Avirulent spirochetes localized in the tissues cause comparatively mild inflammatory reactions. Syphilitic inflammations of this type occur in all tissues and organs; they are most easily recognized in the nervous system, heart, aorta, pancreas, suprarenals and testes.

In the course of time syphilis tends to become a mild process, but at any time the partnership between the body and the spirochete may become disturbed. The tissue susceptibility or virulence then becomes increased, so that the disease again appears above the clinical horizon. Immunity in syphilis depends upon the carrying of the spirochetes; and the syphilitic is a spirochete carrier. The disastrous effects of syphilitic infection usually require a period of years for their development.

Sociological Aspect of the Disease.—There can be no doubt as to the deteriorating effect of syphilis on our population. According to the United States Census of 1900, about 3,000 deaths annually are directly attributable to acquired syphilis. This is about 3 in every 1,000 deaths. The mortality statistics of life insurance companies tell a similar story. The estimated mortality is 133 as compared with 100 non-syphilitics. Acquired syphilis, therefore, decreases one's expectancy of life by about one-third. In the Surgeon-General's reports syphilis headed the list of diseases causing soldiers and officers to become unfit to follow their profession. Statistics both in America and in Europe seem to indicate that from 5 to 20 per cent. of the male population is infected with syphilis. What this means in terms of money losses can hardly be computed. The loss in health and happiness is something that certainly cannot be interpreted in terms of money.

As to hereditary syphilis with its frightful mortality of from 60 to 85 per cent., only one short phrase can be applied: "It is criminal."

RELATION TO MARRIAGE.—A common question and one most difficult to answer is, "How soon may a syphilitic marry without the risk of infecting the mother and her offspring?" We must state the premises so as to enable the conscientious physician to arrive at a conclusion. It may be axiomatically stated that syphilis is a curable disease. Experience has repeatedly demonstrated that patients have passed through syphilis and into old age after having founded large families, the members of which had remained free from the disease during their lifetime. Perhaps another argument in favor of the curability of syphilis may be adduced by mentioning the possibility of re-infection in cured cases, for it is generally admitted that an individual still suffering from syphilis cannot be reinfected. If the curability of syphilis is admitted, we cannot withhold consent to marry from those in whom there is an absence of clinical signs and whose blood and spinal fluid, having been repeatedly examined for Wassermann, were found negative. Certainly no one should be permitted to marry who still suffers from symptoms of the disease, it being quite evident that the wife may be infected through

the patient's secretions and from her own offspring—her syphilitic child. Absence of clinical signs and symptoms does not necessarily prove non-existence of the disease, for there may be long periods of latency. It is necessary to state, however, that the longer the period which has passed between the infection and the contemplated marriage, the less likelihood there is of infecting the mate. At the same time one must remember that the period of the transmissibility of the disease to offspring is longer than the period during which infection may take place. Though the chances of infection are materially reduced after three to four years, yet there are numerous instances of infection having occurred five and ten years after the primary sore. Time, therefore, is not the sole criterion.

Of greater importance is the *type* of disease from which the patient suffers and the *course* it has followed during the first few years after thorough treatment. Merely because a case has run a benign course with little or no treatment is no guarantee that the patient will not later develop interstitial or parenchymatous neurosyphilis. Indeed, experience has taught the opposite—patients with mild early symptoms who had inadequate or no treatment have later become the victims of some form of nervous syphilis, notably tabes and general paresis. A more favorable view may be taken of the robust individual who has observed the best hygienic rules and who has received adequate and continuous treatment during the first three or four years following infection. In the cases in which there have been frequent recurrences of secondary and tertiary lesions in skin and mucous membranes, lasting a number of years, consent should not be given. Certainly in cases of so-called "malignant lues" with deep-seated syphilids and cachexia, patients should be warned against marriage. While one may somewhat hesitate to withhold consent from an individual who only suffered from mild skin lesions, extreme caution should be exercised in advising marriage to those who have suffered from visceral syphilis and neurosyphilis. In the case of frequent recurrences of nervous symptoms, this alone should constitute the most powerful argument against marriage. Our decision must also of necessity be influenced by a knowledge of whether the patient has received sufficient and the proper kind of treatment. The more thorough the treatment has been, the less likelihood is there of infecting one's spouse.

The danger of infection is comparatively slight when symptoms have always been mild, the serological examination repeatedly negative and symptoms have been absent for three years. In such cases the patient may be given additional courses of antispecific treatment and upon the reappearance of symptoms, or when the Wassermann becomes again positive, marriage must be postponed for two more years, during at least one of which there should have been no symptoms even after all treatment has been discontinued. Certainly the existence of parenchymatous, or even interstitial neurosyphilis, forbids marriage.

Classification.—Neurosyphilis may advantageously be discussed under the headings of (1) *Interstitial Syphilis*, to which belong most forms hitherto treated under (a) cerebral, (b) spinal, (c) cerebrospinal syphilis; and (2) *Parenchymatous Syphilis*, among which are placed the diseases formerly grouped under parasyphilis or metasyphilis, namely, (a) tabes, (b) general paresis, and (c) certain forms of progressive muscular atrophy.

INTERSTITIAL NEUROSYPHILIS**(a) CEREBRAL SYPHILIS**

The symptoms of brain syphilis are by no means pathognomonic, for identical disease pictures may be produced by non-specific lesions. Thus, hemiplegia, the result of syphilitic thrombotic processes in the blood-vessels, does not differ essentially from hemiplegia caused by a thrombosed vessel from atheroma. Similarly, localized convulsions caused by meningo-encephalitis or gumma are indistinguishable from convulsions due to tubercle or neoplasm. The numerous syphilitic cranial nerve palsies present a symptomatology identical with that caused by non-specific exudates at the base of the brain. In these and other instances, the study of the particular grouping of symptoms is extremely valuable, for syphilis has peculiarities of its own. Thus, an organic condition which somehow fails to conform to the classical disease picture—a left-sided hemiplegia with aphasia in a right-handed person, ophthalmoplegia on one side with partial paralysis of the opposite one, or incomplete paralysis of one side combined with unusual complications on the other side—compels a careful search for a syphilitic etiology. More suggestive yet of syphilis is the tendency for symptoms to appear and disappear, reappear and perhaps again disappear to become permanent later. A paralysis, having developed on one side, may speedily recede and shortly thereafter make its appearance on the opposite side. An ocular palsy may be discovered in one eye and shortly thereafter in the other. Likewise, temporary blindness of one eye may be followed by permanent blindness of the other, or perhaps of both eyes.

Certain of the symptoms occasionally act as forerunners to more serious ones and may well be styled *premonitory symptoms*. Of these the important ones are: headache, insomnia, vertigo, physical and mental fatigue—symptoms which make their appearance perhaps weeks or months before the complete syndrome of cerebral syphilis is established.

Headache may present itself as one of the earliest manifestations of brain syphilis and always demands careful investigation. The cephalalgia is usually intense and has a tendency to become worse toward evening, or it may be present during the late afternoon and evening hours, though numerous instances are recorded of syphilitic headaches appearing only during the day. In location the headaches may be unilateral or bilateral, frontal, temporal, or occipital; or they may be shifting in character. Such cephalalgias may precede an attack of hemiplegia or aphasia by days, weeks or months; but with the development of paralysis the headaches usually disappear or become less intense. Headaches have been known to be present as the sole symptom of brain syphilis, to which no other symptoms have subsequently been added; out this is uncommon.

Insomnia and restlessness are symptoms commonly found associated with cephalalgia, but may be present without it. *Intractable insomnia* has often preceded the development of other symptoms of brain syphilis.

The very opposite state, namely, attacks of *somnolence* from which the patient can be roused for short periods only, may constitute an early symptom of brain syphilis.

The following premonitory symptoms may be mentioned as occasionally heralding the approach of brain syphilis: vertigo, mental apathy, lack of concentration and loss of memory, inability to find the correct word in ordinary conversation, confusion of speech, epileptiform con-

vulsions, depression or exaltation of spirits. In almost every case of brain syphilis careful search will reveal the existence of some or all of the symptoms enumerated, though patient and relatives may have ignored their presence.

For a detailed description, cerebral syphilis may be discussed under three headings, namely, (i) the vascular or arterial type, (ii) the meningeal variety, affecting either cortex or base, and (iii) the gummatous variety. Combinations are also frequent; thus there are meningovascular, meningogummatous, and meningovascular gummatous types.

(i) *Cerebral Vascular Syphilis*

The usual symptom of this variety is hemiplegia, and if the occluded middle cerebral artery is on the left side, there will be a combination of hemiplegia with aphasia of variable degree. As already mentioned, hemiplegia from syphilitic thrombosis presents pictures differing nowise from hemiplegia of non-specific origin. There are features peculiar to the syphilitic types of hemiplegia. In these the paralysis is rarely attended by loss of consciousness, and warnings are usually present. With the appearance of paralysis the nocturnal headaches, usually present before the attack, either disappear or become less intense. Syphilitic thrombosis is capable of producing two sets of phenomena which may follow each other in more or less rapid succession, namely, *temporary* and *permanent* hemiplegia.

The temporary palsies are probably caused by transient anemia in certain brain territories from gradual and incomplete occlusion of the vessels, while permanent paralysis follows complete occlusion of the arteries. If the circulation continues to improve by means of reëstablished collateral sources, or because of increased heart action, the hemiplegia may improve or disappear entirely. On the other hand, in the event of complete arterial obliteration, there is no prospect for the establishment of a collateral circulation, and the hemiplegia remains complete and permanent. As a result of temporary and incomplete occlusion there may be observed: temporary and incomplete hemiplegia, partial aphasia, mental confusion of short duration, partial loss of consciousness, vertigo, paresthesia and muscular twitchings. Complete blocking of blood-vessels causes a complete and more or less permanent establishment of the conditions just enumerated. In the last alternative it is evident that antispecific medication can produce little or no effect on the disease. One might state as an axiom that brain softening, once it has taken place, remains in spite of antispecific treatment: nothing avails against complete arterial occlusion, produced by whatever cause. In point of frequency the middle cerebral artery and its branches are most often affected in vascular brain syphilis, while the basillar artery with its tributaries ranks next.

As vascular brain syphilis is principally concerned with the immediate and remote effects of thrombosis of the middle cerebral artery—namely hemiplegia and aphasia—a short description of these syndromes

is required. In this connection we can afford to be brief, as the symptomatology of syphilitic hemiplegia and aphasia presents no radical or essential differences from that of the non-specific variety.

Because thrombus formation is necessarily a slow process, *premonitory* symptoms will be present. Warnings usually come in the form of headache, vertigo, dizziness, and transient motor weakness. The motor disability is not mere fatigue; it is a real deficit in the dynamic power of a muscle or group of muscles which the patient feels and the physician may ascertain by objective examination. The weakness may remain minutes or hours; it may express itself in clumsiness of the hands or in a tendency to drop things, perhaps also in a stumbling gait or a dragging toe. These symptoms, transient at first and considered trifling, continue to recur with greater frequency and intensity, and cause the patient much alarm. He no longer mentions his "spells," but dwells upon his "attacks." On the sensory side numbness and formication in the hands are experienced. If the arterial occlusion implicates the left side of the brain, there will soon be added speech disturbances in the form of thick speech or inability to find the proper word in ordinary conversation. Meanwhile the numbness in the extremities continues, no amount of rubbing seeming to relieve the paresthesia or the feeling of "deadness" and "numbness." The fingers may not be able to button the coat so well, and the leg may be dragged considerably in walking. These and similar transient symptoms of disability may occur at intervals during weeks and months until complete arterial occlusion produces the finished picture of thrombotic hemiplegia. As the result of syphilitic arterial disease with a special predilection for the purely psychic centers, mental symptoms may occur in the form of sluggish cerebration and confusion of thought, forgetfulness, falsification of memory, irritability, hypochondriasis, hilarity alternated by crying. Indeed, mental disturbances not infrequently precede or follow the usual attack of luetic thrombosis eventuating in hemiplegia.

The *somatic* symptoms are few in number. There are practically no temperature changes; very rarely is a rise of from 101° to 102° F. (38.3° to 38.9° C.) noted. The unilateral elevation of temperature so characteristic of apoplexy from cerebral hemorrhage is absent. The pulse is soft, compressible, weak and rapid, but may be normal. The paralysis itself may be so slow in its development that at some stage of its progress it may appear as a true monoplegia. Likewise, localized convulsions, so-called jacksonian epilepsy, may appear in this form of brain syphilis, though this syndrome is more common in meningitis of the convexity. Aphasia, when it results from vascular occlusion, may assume any of the well-known types—motor, sensory, or mixed. The character of the aphasia will depend entirely upon the group of vessels affected; the tendency for all forms of aphasia is to become more or less permanent.

Thrombosis of blood-vessels supplying sensory and special sense centers will produce the symptoms characteristic of interference with their nutrition. Thus, tingling and numbness, with varying degrees of an-

esthesia, are the consequences of arterial occlusion of the blood supply to the postcentral convolution. In a similar manner are produced a variety of symptoms too numerous to mention, all depending upon the territory which is impoverished of its blood supply.

Cerebral hemorrhage is rarely caused by syphilis of the brain. In the small percentage of the cases in which this has occurred, there were present aneurysmal dilatations of blood-vessels which ruptured after they had become weakened from syphilitic infiltration. The artery most often affected is the basillar at the base of the brain, and the termination is usually fatal. This artery is seldom attacked by either thrombosis or hemorrhage, except in extreme old age, when the arteries have become brittle. When, therefore, symptoms of basillar arterial disease appear in a young individual, the probable diagnosis is syphilis of the brain.

The following case may serve as a fair example of vascular brain syphilis:

CASE I.—The patient, a woman, aged 30, was brought to the Cook County Hospital for the relief of headache and paralysis. The examination revealed a right-sided hemiplegia with sensory and motor aphasia. The patient appeared to suffer from intense headaches. The onset of the attack was gradual, the right half of the body becoming slowly useless: leg, arm and face, in the order given. At no time during the attack did she lose consciousness. Deep reflexes were exaggerated, Babinski sign was present on the right side. The pupils were irregular and reacted sluggishly to light and well to accommodation. Globulin reaction by Nonne and Ross-Jones positive; Wassermann on blood and spinal fluid strongly positive. We learned that two weeks before the attack intense headaches—worse at night—made their appearance; these became less intense after the complete development of the right-sided hemiplegia with aphasia.

(ii) *Syphilitic Meningitis*

The meninges may be affected over the convexity or at the base of the brain. For clinical purposes the two forms may be discussed under separate headings.

(a) **Syphilitic Basillar Meningitis.**—Headache is an early symptom; it may appear in paroxysms, or it may be more or less continuous. In point of time the cephalalgia is usually more intense at night, though it may persist in the daytime. There are attacks of vertigo, nausea and vomiting, as well as transient losses of consciousness, followed perhaps by convulsions. In the majority of the cases some degree of mental impairment is noted, which may in some instances merge into definite dementia. Memory is poor, and there is a peculiar stupor. It is remarkable that patients who have been comatose or semi-comatose for days and weeks will suddenly awaken as though nothing had happened and, after beginning a conversation, relapse into a state of stupor. To the uninitiated the patient appears to be in deep sleep or in a state of

profound intoxication. The somnolence may be suddenly interrupted by attacks of violent delirium, or a state of mental confusion may supervene. The patient appears to have the extraordinary ability to emerge from delirium into coma, and from deep stupor he may as readily return to perfect lucidity.

The symptoms thus far enumerated may well be called the *general symptoms* of syphilitic meningitis. It is now necessary to mention the so-called *localizing signs* of syphilitic infiltration due to the formation of exudates at the base of the brain. These consist mostly of paralyses or palsies of the several cranial nerves situated at the base of the brain.

Owing to the frequent localization of meningitic processes at the interpeduncular space, the optic chiasm and ocular nerves are often affected. Involvement of the optic chiasm in its anterior portion gives rise to bitemporal hemianopsia, that is, blindness in both outer fields of vision, because the exudate compresses the innermost portion of the optic chiasm containing the visual fibers for both outer fields. This is the well-known syndrome of hypophysis tumor, which may be reproduced by syphilitic basal meningitis.

The cranial nerve most frequently affected by the luetic basal disease is the oculomotor or third nerve, which is compressed by a meningeal exudate. That oculomotor disease is not necessarily the result of gumma, is evidenced by the clinical observation that the paralysis is usually transient, capable of complete regression. Further, rarely is there *complete* third nerve paralysis; most commonly there is only *partial* paralysis of one or more branches. The abducens or sixth nerve may be affected on the same side as the third nerve.

Associated with nerve palsies are usually headache, vomiting, vertigo, partial paralysis, convulsions, optic neuritis, and the other signs and symptoms of basal meningitis. However, involvement of a single branch of an oculomotor nerve may be the sole symptom of a basal process. The palsy may be slight, consisting perhaps of an insignificant ptosis, or a tendency to strabismus. Indeed, the palsy may be so transient that, when looked for on the day following its appearance, every trace of it may have disappeared. Transient palsies not only have diagnostic value, but are also of prognostic importance: their appearance usually indicates that more trouble may be expected. Quite frequently one branch after another becomes paralyzed because of the gradual cutting off of the blood supply, owing to progressive meningeal infiltration.

In the same way the trigeminus nerve may be affected alone or in association with other nerves. Even the gasserian ganglion may be caught in the luetic process. The symptom will be trigeminus neuralgia, followed by anesthesia of the trigeminus. There may be a combination of trigeminus involvement, disease of adjacent cranial nerves and hemiplegia.

With localization of the meningitis on the side of the pons, we expect palsy of the fifth, sixth, and seventh nerves, associated perhaps with hemiplegia of the opposite side. Rarely is the motor branch of the

trigeminal nerve attacked; the symptoms will then be those of paralysis of the muscles of mastication.

Seventh nerve paralysis is not uncommon and usually occurs in combination with auditory paralysis. In fact, a combined unilateral facial and auditory palsy, unless caused by trauma, always suggests syphilis.

The vagus, glossopharyngeal, and hypoglossal nerves, when the seat of syphilitic deposits, give rise to symptoms which are identical with those found in the non-specific types. There will be palsy of the soft palate, paralysis of the vocal cords, and atrophy of the tongue.

Should the spinal accessory nerve become involved in theluetie infiltration, there will be paralysis of the sternomastoid muscle and of the upper portion of the trapezius.

In basal meningitis, almost any combination of lesions, with their corresponding symptoms, is possible. For instance, paraplegia may be the result of disease over the pyramidal decussation, and cerebellar symptoms may appear in consequence of a syphilitic exudate over the cerebellum.

Certain features of this type of brain syphilis are illustrated by the following two cases from the writer's hospital service.

CASE II.—The patient, a laborer, 29 years of age, entered the hospital for the relief of headaches which were intense enough to prevent sleep. He was extremely apathetic; would begin a conversation, say a few words, and then relapse into a stupor resembling that of profound alcoholism. The headaches, which were always worse afternoons and evenings, began about three weeks before admission. The objective examination revealed exaggerated tendon reflexes, but no Babinski, Oppenheim or Gordon signs; there was no ankle clonus. While none of the voluntary muscles showed evidence of paralysis, speech was thick and indistinct. The eye-grounds revealed typical optic neuritis.

CASE III.—The patient, a washerwoman, 40 years of age, has been married 25 years. She is the mother of five living children, there were three miscarriages. The present trouble seems to have had its beginning about two years ago, when she developed severe headaches which have been almost continuous, though there were remissions and exacerbations. About eight months ago there appeared peculiar paresthesiæ—crawling sensations over the entire left side of the face. Six months ago strabismus and diplopia were added to the other symptoms. Under energetic antispecific treatment by means of mercury and iodids, the ocular palsies causing diplopia and strabismus had disappeared, but two months ago the patient discovered that her face was drawn to the right. There was a well-marked left-sided facial paralysis. The pupils were extremely irregular, but showed no Argyll Robertson phenomenon. To summarize: there were tingling and numbness in the left half of the face—trigeminal involvement. Somewhat later, ocular palsies appeared, from which there was complete recovery. Toward the last, evidences of

left-sided peripheral facial paralysis were discovered. The Wassermann test on blood and spinal fluid was strongly positive.

(b) **Syphilitic Meningitis of the Convexity.**—In this type of luetic meningitis the principal symptoms are referable to involvement of the cortical centers. Thus, there may be monoplegia, hemiplegia, aphasia—motor, sensory, or mixed. Perhaps the most important symptoms are localized or general convulsions with transient palsies. To these are usually added the general manifestations of basal meningitis.

(iii) *Gumma of the Cerebrum*

The symptoms of gumma of the brain are similar to those produced by a non-specific tumor. There are (1) the general symptoms of brain pressure, consisting of headache, nausea and vomiting, choked disk, general malaise and vertigo; and (2) the local symptoms indicative of the exact portion of the brain which is being compressed, differing with the location. The so-called specific features which distinguish syphilitic from non-syphilitic tumors are only suggestive, there being no pathognomonic differential signs. The course and localization of the disease may be of assistance in differentiation. In syphilis the course is mostly subacute or subchronic and the localization in the brain is superficial. It may appear at either end, vertex or base. Only occasionally has it been seen in the pons and but rarely has gumma been discovered in the cerebellum. Owing to its localization in the cortex, a gumma is more likely to produce symptoms of cortical irritation than is a non-specific tumor. This may explain the greater frequency of convulsions in gumma, and their relative rarity in non-specific neoplasms. Localized convulsions beginning with an aura may give important clues as to the localization of the process. After everything has been said on this subject, the fact remains that all symptoms observed in specific tumor may also be seen in the non-specific variety. It is in just such cases that the Wassermann test becomes of immense diagnostic importance.

The following is an abstract from the history of a case illustrating this variety of brain syphilis.

CASE IV.—The patient, a man 40 years of age, entered the hospital for the relief of headache, nausea and vomiting, accompanied by poor vision and general weakness. The symptoms had been developing for the past two months, becoming progressively worse and vision constantly lessening. The objective examination revealed choked disk with beginning secondary optic atrophy. The pupils were unequal, the left being larger than the right, and neither reacted to light, but responded promptly to accommodation. Under treatment with mercury and salvarsan the headaches disappeared, the swelling in the disk was reduced, but the secondary optic atrophy became more marked. The picture in this case was of both tumor and gumma. Only careful observation,

results of treatment, and a positive Wassermann made the diagnosis—gumma of the brain—a certainty.

As previously stated, neurosyphilis rarely limits its activities to one tissue or to one location. In the same patient there may be present manifestations of gumma, meningitis and even endarteritis. Usually one of the three types presents the leading symptoms, while the other varieties may indicate their presence by a number of less important symptoms. Likewise, the disease may appear simultaneously in several locations, or it may invade several regions in quick succession.

Intellectual disturbances are seldom absent in interstitial syphilis. The frontal lobes are not usually the seat of syphilitic disease, but they do not always escape invasion. The mental syndromes of frontal lobe involvement may resemble almost any of the well-known types of insanity, especially mania and melancholia. The most important intellectual deficit is that of so-called dementia on a syphilitic basis. This disturbance is not to be mistaken for parietic dementia, or general paresis—a parenchymatous form of syphilis to be described later. The symptoms of syphilitic dementia—an interstitial type of neurosyphilis—are so similar to those of general paresis that differentiation in some instances is extremely difficult. This form of interstitial syphilis has also been called pseudo-dementia syphilitica. Possibly many of the wonderful "cures" of general paresis were effected in individuals who suffered from this type of disease.

(b) SYPHILIS OF THE SPINAL CORD

The spinal cord, like the brain, may be affected by syphilitic processes either in its blood supply or in the cord substance proper. It is convenient to speak of syphilitic myelitis, meningomyelitis, and Erb's spinal syphilis.

The symptomatology of syphilis of the cord varies according to the extent, intensity, diffuseness and location of the lesions. There are certain symptoms suggestive of syphilis, while the majority of symptoms are those found in spinal cord diseases, regardless of etiology. There is something unfinished or rather incomplete in the picture of syphilis of the cord, while non-specific cord disease usually conforms to a type and often corresponds to textbook description.

As the majority of syphilitic cord cases have their earliest pathology in the meninges, pain in the back is a prominent symptom. This is usually worse at night—a characteristic of all syphilitic pain. Further, as the disease progresses, more or less of the cord substance proper becomes implicated, and we then have the symptoms indicative of cord involvement, namely, anesthesia, paralysis of muscles and sphincters. Again emphasis must be placed on the clinical fact that the anesthesia as well as paralysis may be incomplete and transient, and may come and go several times before it finally becomes complete and more or less permanent.

As the meninges become surrounded by syphilitic exudate, the posterior roots passing through them are irritated. The result will be severe pains radiating into their corresponding nerve-trunks, girdle pains and neuralgiform pains in the extremities. In severity and persistency the pains vary from almost unbearable and continuous suffering to mere light transient twinges of pain.

Paralysis of motion may be of the upper motor neuron variety, with spasticity, exaggerated reflexes, and the utter helplessness of spastic paraplegia; or the patient may present a mere clumsiness in walking, especially in ascending a flight of stairs. In this form of paralysis there will be no wasting of muscles and no electrical changes. When the anterior horns become the seat of the disease, there will be rapid atrophy of muscles, loss of the reflexes, and a flaccid type of paralysis. As the dorsal or thoracic portion of the cord is the one usually affected by syphilis, atrophy of intercostal muscles must occur. These are not easily recognized, because the symptoms are so indefinite.

The important symptoms of luetic cord disease are to be found in the lower extremities and depend largely upon the extent and portion of cord affected. It is quite possible for one-half of the cord to become diseased and to present the picture of a Brown-Séquard paralysis. The motor paralysis will then be found on one side and the sensory disturbance will be principally limited to the opposite side. The syndrome is usually incomplete from the beginning, and if complete, does not remain so for any length of time.

To the motor and sensory disturbances in the lower extremities there are soon added sphincter troubles. The bladder may at first act capriciously: there may be either frequent urination or difficulty in expelling the urine. This may soon pass off, or the difficulties become more accentuated. The patient then suffers from incontinence and retention. In a similar manner the rectum may show signs of incompetency by developing obstinate constipation, or by the inability to retain its contents after cathartics have been administered. Retention rather than incontinence is the rule with the rectal sphincter. The sexual functions may also show impairment—there may be premature ejaculations or impotence, may be complete, though desire may still be present. Like the other syphilitic manifestations, sphincter disturbances may be of all grades of severity and may be extremely evanescent.

Spinal cord syphilis includes not only disease of the cord proper, but also meningeal and root syndromes which may appear singly or in combination. In this respect it resembles brain syphilis, which presents numerous combinations; indeed, brain and cord syphilis so commonly appear together in the same individual that the term cerebrospinal syphilis has been coined to express this combination of symptoms.

Among the several types of cord syphilis, the group described by Erb must receive special mention. There is considerable doubt as to whether the symptoms are sufficiently distinctive to entitle the syndrome to be placed in a group by itself, but we shall follow custom and discuss it separately.

In *Erb's spinal syphilis* the development is slowly progressive and usually begins with spasticity of a light degree, but with considerable clumsiness in locomotion. After slight exertion the patient experiences a sense of fatigue, out of all proportion to the effort put forth. Especially difficult is it for him to ascend a flight of stairs and to overcome slight obstructions in his path. A frequent complaint, for instance, is the patient's inability to step on elevated ground without falling. The disease itself, very chronic in its evolution, may show remissions, and the patient may not find it necessary to consult a physician. The sensory symptoms are slow in coming, and when present, are rather insignificant; sphincter troubles come late in the disease and share the tendency of other syphilitic manifestations to appear and disappear when least expected. The tendon reflexes are exaggerated and the pathological reflexes, such as Babinski's sign, Oppenheim and Gordon phenomena, are usually in evidence.

Spinal cord syphilis may yield symptoms resembling tabes dorsalis when the posterior columns suffer the brunt of the syphilitic attack. We may then find nearly all the cord symptoms which we are accustomed to see in tabes. There should be no difficulty in differential diagnosis when careful search is made for symptoms other than frank cord symptoms. These cases have been described under the term *pseudo-tabes syphilitica*.

Though the mid-dorsal cord appears to be the most common site of syphilitic cord involvement, any portion of the spinal cord, and even the medulla, may be the seat of gummatous deposit or meningitic infiltration. If the disease is in the upper part of the cord, we expect symptoms of cervical cord involvement which do not differ from those caused by other factors. In cauda equina disease caused by syphilis there will be produced the syndrome of cauda equina lesions—partial paralysis of the lower extremities, radiating pains in the region of the lumbosacral nerves, partial anesthesia in the equina roots, and bladder, rectum and sexual disorders.

Reflexes.—The condition of the deep reflexes will vary with the particular portion of the cord involved. The rule is, that the reflexes are increased in cervical, dorsal and upper lumbar involvement, while a decrease or loss occurs when the lower lumbar, sacral cord or the cauda equina roots become the seat of disease.

Motility.—The motor paralysis is of the upper motor neuron type when the cord is affected above the second lumbar segment, of the lower motor neuron variety when the parts diseased are situated below that segment. It may not be amiss to reiterate what is meant by upper and lower motor neuron paralysis. We understand by *upper motor neuron paralysis* that form which is characterized by spasticity, exaggerated reflexes, the presence of Babinski sign or its equivalents, and the absence of muscle atrophy and of electrical changes. On the other hand, *lower motor neuron disease* is known by the fact that the muscles are wasted—atrophic—reflexes are absent, the paralysis is of the flaccid

variety and the well-known electrical changes of reaction of degeneration are present.

Sensation.—The sensory disturbances encountered in spinal syphilis are not only those of irritation—namely, sharp, darting pains radiating into the extremities—but also of destruction: numbness, paresthesia, anesthesia and analgesia of varying intensity. The degree of sensory disorder will depend largely upon the amount of pressure exerted by the syphilitic exudate. Sensory syndromes similar to those of syringomyelia have been observed; the pain and temperature senses have been abolished or reduced, while tactile sensation has remained unimpaired. The reverse condition is also possible, viz., tactile sense may be lost, while pain and temperature remain normal.

Trophic Changes.—Not only is there degenerative atrophy of muscles when the anterior horns are affected, but in almost all forms of spinal cord syphilis there is a tendency to the formation of bed-sores and deep ulceration of skin.

Sphincter Paralysis.—In practically all varieties of syphilis of the spinal cord the sphincters are paralyzed either partially or completely. It must be emphasized that this tendency is more marked in non-syphilitic cord disease and may serve as a differential point in diagnosis.

To better illustrate the symptomatology of spinal syphilis, the writer will reproduce, in part, a clinical lecture on spinal syphilis with presentation of cases:

CASE V.—The patient, an Austrian shoemaker, widower, 52 years of age, complains of weakness in both legs and inability to walk. About six years ago he noted that his legs were gradually getting stiff and clumsy, but he at no time experienced sharp pains. There were times when he thought he was getting better, but on the whole the disease has been progressive. During the last few months he noticed bladder disturbances in the form of partial retention, also rectal trouble in the form of obstinate constipation. Gradually the legs became weaker, until complete paralysis appeared about three months ago. He is now unable to leave his invalid chair. An objective examination reveals extremely exaggerated reflexes in the lower extremities, Babinski sign and ankle clonus are marked. There is present slight sensory disturbance over the legs and feet, affecting principally the deep sensibility. He denies venereal infection, but admits exposure for a period of 15 years, the time during which he has been a widower. The pupils are unequal, barely respond to light but well to accommodation; in other words, they present an incomplete Argyll Robertson phenomenon. The Wassermann is positive on the spinal fluid and negative on the blood; there are 50 lymphocytes per cu.mm., and the Nonne globulin test is strongly positive. Diagnosis: Spinal syphilis of the Erb type.

CASE VI.—The patient, a teamster, unmarried, and 30 years of age, entered the hospital because of stiffness in the legs. An examination demonstrates the presence of exaggerated knee jerks and brisk Achilles

reflexes, with Babinski sign on both sides. In addition, there is a genuine ankle clonus. When the feet are approximated and eyes closed a typical Romberg sign develops—the patient's body sways. In isolated areas there is found anesthesia and analgesia on the lower extremities, but no distinct level of anesthesia. The tuning-fork test reveals a disturbance of the osteo-sensibility. There is evidently not only disease of the pyramidal tracts, but also of the posterior columns. The patient denies syphilis and even exposure. As regards the beginning of his trouble we elicit the information that he experienced intense pains radiating down his legs at first; these gradually disappeared, but recurred several times. When paralysis appeared the pains did not return. Evidence of a syphilitic etiology are the unequal pupils, which are rigid both to light and in accommodation. Wassermann is positive. The diagnosis is syphilitic cord disease of the meningomyelitic variety. The pathological changes were in the meninges first; this gave rise to the root pains. Later the cord was compressed and the pyramidal tract as well as the posterior columns became affected.

CASE VII.—The patient, a married woman 32 years of age, complains of jerking and twitching in both legs, inability to walk, and involuntary urination. The disease began three months before admission to the hospital with sharp pains radiating down the legs and into the abdomen. At the same time there appeared occasional contractions and twitchings of the leg muscles. Both legs were not affected at once, one preceded the other; soon both extremities became weak, and a month ago complete paralysis and stiffness developed. The involuntary action of the bladder became gradually worse, until complete incontinence remained. The examination reveals negative findings in the upper extremities, but all power is lost in the lower extremities. The muscles feel flabby and the deep reflexes are lost in the lower extremities. The Babinski toe-sign is present on both sides. There is anesthesia and analgesia over both lower extremities extending up to the umbilicus, but leaving out small areas in an irregular manner. There is a massive bed-sore over the back, extending from just above the rectum to the sixth dorsal vertebra, laying bare the back muscles to a depth of at least five inches from the surface. Though the patient denies syphilis, the positive Wassermann makes this diagnosis definite. We assume in this case involvement of the lumbosacral cord, because the reflexes in the knees and in the Achilles tendons are lost, the paralysis is a flabby one, and the sensory deficit is limited to the lumbosacral cord. But there can be no doubt as to the dorsal cord disease, for the Babinski sign would not be possible without an upper motor neuron affection. The question arises as to the possibility of obtaining a Babinski phenomenon when the lumbar cord is destroyed. We must assume that the pathway from the upper portions of the cord is not entirely interrupted, that we are dealing with an incomplete myelitis. This is also supported by the finding of a few irregular areas on the lower extremities in which sensation is not destroyed. The case appears to be one of diffuse myelitis involving

principally the lumbar and sacral cord and to a certain degree also the dorsal cord.

Having perused these reports of cases of true spinal syphilis, the reader is now prepared to understand an account of so-called cerebrospinal syphilis, by far the most common form of interstitial syphilis of the nervous system.

(c) CEREBROSPINAL SYPHILIS

In cerebrospinal syphilis there are present brain symptoms *and* spinal cord symptoms in various proportions, sometimes one set preponderating, sometimes the other. The patient may have suffered from an attack of hemiplegia or from a cranial nerve palsy and later develop an attack of spastic paraplegia with urinary and rectal sphincter incontinence. This is clearly spinal cord disease superadded to a cerebral affection—a cerebrospinal syphilis. Or, the spinal cord symptoms may have preceded the cranial symptoms, in which case we also have cerebrospinal syphilis, though the spinal symptoms preceded the others. We thus get the most varied combination of symptoms, making description of types an impossibility. The writer believes more will be gained from the citation of a few representative cases which follow:

CASE VIII.—The patient, a bookkeeper, aged 32 years, complains of difficulty in walking and loss of sensation in the legs. There are also urinary incontinence and absolute constipation. The disease began gradually about two years ago. He first noticed that he was not sure of himself when getting off and on street cars. Difficulties in walking became more pronounced later. One evening while at a card party the cards dropped out of his hand which he felt becoming weak and lifeless. The following afternoon he suddenly fell to the floor, the leg weakness, from which he had practically recovered the day previous, having returned. This condition has remained stationary. Quite recently marked urinary and rectal incontinence has appeared. One year ago he suffered from intense occipital headaches lasting over a period of several months. A few months ago he had an attack of speechlessness lasting a few minutes, during which he could not articulate plainly—could not pronounce his words well and was not understood. His family and personal history is negative. He knows of no syphilitic infection, admitting only a case of gonorrhea eleven years ago. The *objective* examination reveals a slight inequality of the pupils, which react to light and in accommodation, though the right pupil is somewhat sluggish to light. The kneejerks are both exaggerated and there is well-marked ankle clonus and double Babinski sign. The legs are spastic and the picture is that of a true spastic paraplegia. There is a reduction in tactile and pain sense of the entire lumbosacral territory, beginning opposite the first lumbar segment. In addition, there are retinal hemorrhages in the right eye, which caused the patient to become partially blind in that eye.

In this case some of the symptoms are so definitely cerebral that no doubt can be entertained of their genuineness; and at the same time the remaining symptoms are of distinctly spinal character. The combination is cerebrospinal syphilis, because the patient subsequently admitted syphilis and the laboratory proved it by the finding of a positive Wassermann in the spinal fluid and blood.

CASE IX.—Patient is a business man, 45 years of age, who has difficulty in walking and has urinary troubles. He admits having had a chancre 15 years ago, for which he received energetic mercurial treatment during a short period. About 10 years later he developed frequent nocturnal headaches and at times spells of insomnia lasting a week or more. Shortly after the recrudescence of symptoms pointing toward syphilis, he suffered from left-sided ocular ptosis and right-sided hemiplegia, both of which were incomplete and disappeared after two months' treatment. He had frequent reminders of his disease in the form of transient attacks of pains and occasional numbness in the lower extremities occurring at irregular intervals. He received irregular treatment, but not the energetic combined mercury and salvarsan courses which he should have had. About four months ago his legs became numb and cold, and a little later, weak and easily fatigued. At about the same time there were difficulties in voiding urine and in the rectal functions. These symptoms continued to increase until the leg paralysis became more complete. At no time did he experience the sharp pains in the lower extremities so often noted in meningomyelitis. An *objective* examination reveals unequal pupils in which there is no light reaction, though the optic nerve is not involved. No trace of ptosis is to be found. There are no asymmetries of the face and the tongue protrudes in a straight line. The reflexes are pathologically exaggerated, on the right side more than on the left—a remnant of right-sided hemiplegia. The lower extremities are spastically rigid and of the lead-pipe variety. He is barely able to move about with the aid of two crutches and is generally helpless. The sphincters have not given him as much discomfort of late as formerly. Sensation in the lower extremities is markedly impaired in all qualities, but there is no complete anesthesia or analgesia; the symptom is best called hypesthesia and hypanalgesia. The sexual functions are lost. This represents perhaps as typical a case of cerebrospinal syphilis as one may encounter in neurological practice, the majority of cases being somewhat atypical. The diagnosis offered no difficulties, there being both a history of the infection and the serological findings of syphilis.

PATHOLOGICAL ANATOMY OF INTERSTITIAL NEUROSYPHILIS

As in the clinical discussion, the pathological changes occurring in neurosyphilis will be described under the following subheadings: (1) Syphilitic new formations, gumma; (2) Chronic inflammations, meningitis; (3) Disease of blood-vessels, syphilitic endarteritis of the cerebral vessels.

Because the pathological changes in these three varieties of neurosyphilis affect the interstitial connective tissues, the entire group is now known as interstitial neurosyphilis. This is in opposition to what occurs in another group of syphilitic disorders, of which tabes and general paresis are the leading representatives, in which the pathological changes are found in the parenchyma of cells and fibers of brain and spinal cord.

(1) **Gumma.**—This syphilitic new formation, first described by Virchow, usually appears in small and large isolated multiple swellings, though it may occur in diffusely infiltrating masses.

Macroscopically, gummatus tumors are seen to vary in size from that of a grain of barley corn to a walnut, though very large tumors have also been described. The latter, however, have been considered as mere conglomerations of small ones. In the fresh state gummata appear grayish-red or have a yellowish tinge, changing to a decided yellow or yellowish-white appearance in the regressive state. A fresh gumma gives to the palpating finger the impression of a glue-like or gummy substance—hence the name gumma has been given to the entire group. Succeeding these regressive changes a gumma may become soft, though this is not the rule; the more common change is the transformation of a gumma into a caseous mass, unless a mixed infection causes it to become of the consistency of pus.

As to location, the most frequent seat of gumma is in the dura mater; next in frequency is the convexity of the cortex and the base of the brain. In the last situation gummata commonly select the neighborhood of arterial trunks. Of the brain substance itself, gummatus tumors select the cortex, preferably the region of the central convolutions. They are found, in order of frequency, over the frontal, parietal, and temporal, rarely over the occipital lobes. While gummata are occasionally observed in the cerebellum and pons-medulla, they are rather infrequent in these situations as compared with the cerebrum. Rarely a gumma may localize itself in the hypophysis, when symptoms of hypophyseal tumor are produced.

Gummata rarely present smooth surfaces—they are almost always uneven and adhere to the surrounding tissue, thus becoming indistinguishable from inflammatory exudates in the meninges and blood-vessels. It is quite common, however, to find gumma associated with localized meningeal inflammation, so-called gummatus meningitis.

When a gumma is sectioned, a yellowish homogeneous mass is revealed, surrounded by grayish, more or less translucent tissue, varying in consistency from soft to semisolid.

Microscopically, gummata show the appearance characteristic of granulomata—numerous granulation cells are found at the periphery with a sprinkling of spindle- and star-shaped cells, as well as new vascular connective tissue. Next to the periphery, the inner portions are made up of connective tissue and cells undergoing regressive changes, while the center of the gumma is the seat of numerous necrotic foci appearing as homogeneous opaque masses—the cells and nuclei being poorly

stained. Even giant and epithelioid cells may be encountered, thus making the resemblance of syphiloma to tuberculoma more complete.

Syphilomata contain not only caseous and necrotic areas mixed with granulation cells, but new fibrous connective tissue is also seen to permeate the gumma, which is probably the cause of the shrinking of its various parts, giving rise to its uneven surface.

A gummatous tumor usually begins its growth in the connective tissue of the meninges or arteries, not in the nerve elements themselves, though the latter may become involved. The vessels situated in a gumma show round-cell infiltration, especially in their outer and middle coats; the perivascular spaces are widely dilated and filled with round cells; likewise the new capillaries. Secondly to the interstitial involvement, the parenchyma itself, that is, the nerve-cells, suffer. The cells lose their normal configuration and shrink in size and number. The cell-protoplasm is diminished, the Nissl bodies become disintegrated and only the cell nucleus offers resistance to destruction for a time, but eventually it also undergoes atrophy and dies.

In addition to the vascular connective tissue, the glia, which constitutes the proper supporting connective tissue of the brain, undergoes similar destructive changes.

Summarizing the microscopical appearance of the tissues in which a gumma is localized, there is found an encephalitis with inflammation, atrophy and sclerosis, which changes were primarily produced by the development of numerous gummatous nodules.

(2) **Meningitis.**—Syphilitic inflammation of the meninges may occur simultaneously in all three membranes, or it may begin separately in the dura or pia. When the inflammation affects the dura alone, it occurs subsequent to disease in the bones or periosteum.

When the meninges are primarily diseased, the pathology is really a gummatous process which infiltrates the membranes, or it may be the result of miliary gummata scattered throughout the membranes. Both macroscopically and microscopically gummatous inflammation does not differ from the previously described isolated gummata and their infiltrations of the brain substance. With gummatous meningitis there is almost invariably present a fibrous hyperplastic meningitis. The dura is much thickened and adherent to the soft membranes, pia and arachnoid, all of these appearing glued into one mass.

In the majority of cases meningitis begins in the leptomeninges at the base of the brain, even when all the membranes are involved. There are instances, however, in which the syphilitic process is entirely limited to the soft membranes. Rarely the arachnoid is the only membrane implicated. Usually the soft membranes are intimately adherent, thickened and infiltrated, and give the appearance of one inflamed membrane.

Inflammation and neoplastic formation are so intermingled in syphilitic meningitis that it may be difficult to ascertain which of the two processes predominates. The rule is, the more acute the inflammation, the less marked the neoplastic formation.

Like the picture described for gumma, the dura and the soft mem-

branes present caseous foci, which are recognizable by the fact that they stain poorly, in contrast to normal tissue which stains well.

Because of their intimate connection with the superficial layers of the brain, the meninges cannot be removed from the brain without tearing its substance. This finding might explain the clinical observation that a true meningitis is rather rare—most cases belong to meningo-encephalitis.

While syphilitic meningitis may affect the entire brain—convexity and base—the rule is that the process involves either the convexity or the base, but the most frequent seat of syphilitic meningitis is at the base of the brain, and only few cases of cerebral syphilis have been observed with entire freedom from basal meningitis.

Years ago our attention was directed to the great preference of syphilitic meningitis for the interpeduncular space in the region of the optic chiasm, which explains the great frequency of symptoms from meningeal exudates on the part of the ocular nerves and of the optic nerve. The meningeal inflammation also selects the region of the middle cerebral artery, which explains the apoplexies resulting from occlusion by the meningeal infiltration. It is characteristic of syphilitic meningitis that it may appear localized in a small spot at the base of the brain, so that it is possible for symptoms to appear on part of the oculomotor nerve or only on the optic nerve. Cases have been reported in which a syphilitic meningitis was limited to the trigeminus and the gasserian ganglion. In fact, any one nerve may be injured at the base of the brain by pressure from an inflamed and infiltrated membrane. Likewise, the damage may be done to the vessels which may be compressed, or the vessel walls may become involved by the gummatous process in its contiguity. Even the nerve-trunks themselves may be infiltrated by gummatous deposits, or they may suffer from mere compression. The frequency of cranial nerve lesions and apoplexies from brain syphilis finds its explanation in the preference of meningitis for the base of the brain, where the large arteries and cranial nerves are found.

(3) **Vascular Disease.**—Vascular disease of the nervous system by lues may produce paralysis either mechanically, by meningeal thickening or by gummatous tumors. There may be compression of the arteries from without, giving rise to thrombosis, or the arteries themselves may become infiltrated with gummatous deposits. The most important pathological change in the artery is luetic infiltration of its coats, principally of the adventitia, but also of the media, which is pushed into the lumen of the vessel, causing irregular thickenings and subsequent thrombosis formation. Endarteritis luetica is a disease occurring in young syphilitics, and belongs to interstitial syphilis, while another form of arterial thickening, syphilitic arteriosclerosis, occurs in advanced age and should not be confounded with syphilitic endarteritis.

The pathological changes of neurosyphilis may be summarized in the following paragraphs:

(1) The arteries may be involved mechanically by thickening of the meninges.

(2) The arteries may become diseased by contiguity to syphilitic gummatous or inflammatory processes.

(3) The vessels may become affected by the spirochetes attacking the vasovasorum, with subsequent disease of the adventitia, media and intima. This is the syphilitic obliterating endarteritis described by Heubner.

(4) Genuine gumma may be found imbedded in the arterial wall, giving rise to secondary arterial changes.

(5) Syphilis predisposes the subject affected to atheromatous changes, indistinguishable from non-specific arterial disease.

The consequence of arterial changes is a narrowing of the lumen, followed by complete or incomplete thrombosis. The part of the brain depending for its blood supply on such vessels becomes anemic and later undergoes necrosis with death of the brain tissue. As the arteries principally affected by the luetic disease are usually terminal or end arteries, the basal ganglia and the internal capsule suffer most, and thus is explained the great frequency of syphilitic hemiplegia as the clinical expression of syphilitic arterial disease.

The greatest progress in the pathological anatomy of syphilis was made when Schaudinn and Hoffmann discovered the *Spirochæta pallida*, and Roux and Metchnikoff succeeded in transmitting the syphilitic virus from the human to animals. And though the biology of the spirochetes has not yet been definitely worked out, we have learned something of their mode of affecting the organism. It is immaterial whether the spirochetes themselves or their toxins or endotoxins are the cause of the inflammatory processes. We have learned that the *Spirochæta pallida* are distributed throughout the body by the blood-current and the lymph stream. From the blood of a syphilitic taken three weeks before the development of secondary symptoms, Hoffmann was able to reproduce a local syphilitic lesion in the ape, from which lesion he subsequently derived the typical spirochetes. Neisser was able to inoculate other animals successfully from the primary infection of the lower apes as early as five days after the primary inoculations. Within the short period of fourteen days after inoculation the spirochetes had invaded the entire hemapoietic system, spleen and bone marrow. While distribution takes place along the blood stream, the lymph channels constitute the more common route of distribution of the organisms.

The clinical phenomena of the disease seem to demonstrate the fact that the lymphatic apparatus is early affected, perhaps within a few days after infection. As previously stated, the territory of the optic chiasm and the interpeduncular space constitute the favorite seat of syphilitic involvement, a veritable lake of lymph. The real explanation may be that the base of the brain is permeated by many blood-vessels and also bathed profusely by the lymph stream, both of which systems carry spirochetes and offer a favorable nidus for their settlement. It thus comes about that the cranial nerves are frequently affected by the syphilitic poison. As may be recalled, the subarachnoid space is connected through the foramen of Magendie and the lateral

apertures of the fourth ventricle with the ventricles of the brain. The cerebrospinal fluid being again taken up by the lymph vessels which surround the peripheral nerves of the brain and spinal cord, the optic and oculomotor nerves are apt to become affected early.

Ever since Moore and Noguchi discovered the spirochete in the brain of paretics, we also know that this organism is the direct cause of parenchymatous disease of the nervous system. It may now be stated without fear of contradiction that the *Spirochæta pallida* is the sole cause of syphilis in whatever form it may appear. We have long known that the usual location is in the intracellular lymph spaces, but we now know that they are also found in the nerve-cells. Indeed, from the pathological and anatomical studies made by Warthin, of Ann Arbor, it has become evident that the pathological states produced by the spirochetes are much more frequent in the internal organs than we had hitherto believed. By the Levaditi method Warthin has been able to stain spirochetes from almost all organs of the body in cases of syphilis which have not shown clinical symptoms of the disease during life, and certainly in those who have had manifestations of the disease. Spirochetes have been found in the arterial coats, predominatingly in the adventitia, where they have been encountered in large numbers occupying the external layers of the lymph sheaths, less frequently in the muscularis, external to the elastic membrane, and very sparingly in the intima of the vessels. In the large vessels they are most frequently seen in the vasovasorum, which contain the spirochetes in their walls. It is probably because the blood-vessels of the brain down to the smallest ramifications are surrounded by the vasovasorum and peri-arterial lymph sheaths that syphilitic disease affects these areas.

The arachnoid shows proliferation of connective tissue with collections of lymphocytes and plasma cells and but few leukocytes. The small nodules in the pia mater in the vicinity of the vessels are new connective-tissue cells, principally made up of enormous collections of lymphocytes. The lymphocytosis may be the result of the predilection of the spirochetes for the lymph stream.

PARENCHYMATOUS NEUROSYPHILIS

(a) TABES

This disease is undoubtedly the most common organic disease of the spinal cord and belongs to the group of parenchymatous neurosyphilis.

Etiology.—Formerly much space was devoted to a discussion of the various causes of tabes. While it was recognized by men like Erb, Gowers, Strümpell and others that tabes as a disease was caused by syphilis, it was not known until recently that syphilis is the direct and sole cause of this disease. In other words, a degeneration of the posterior columns of the cord is not secondary to gummatous inflammation of the spinal cord, but is a direct result of the syphilitic virus. The recent

investigations of Widal, Plaut, Nonne, Noguchi, and others have confirmed the theory of the syphilitic origin of the disease. Nonne's modern work on syphilis of the nervous system has proved that all cases of syphilis give a positive reaction in the blood—60-70 per cent.—and that only 5-10 per cent. of spinal fluids give a positive Wassermann reaction with the original method when 0.2 c.c. of fluid is used. With larger quantities, the Wassermann reaction is positive in 100 per cent. of cases. The writer believes that the positive proof of the syphilitic etiology of tabes was rendered when the spirochetes were found in the spinal fluid and in the cord structures.

The disease occurs more frequently in males than in females and in the better classes of society. It is also seen more frequently in the intelligent than among the ignorant; oftener in the Caucasian than in the colored race.

From the time this disease was discovered, in 1861, to the present the following contributory factors have been recognized: exposure to cold; frequent and prolonged fatigue; sexual excesses; intemperance in the use of alcohol and tobacco; infectious disease and trauma. It cannot be denied that as contributory factors some weight must be given to each of these. All investigators have agreed that tabes is a syphilitic disease and that other contributing causes are not to be seriously considered. In those cases where no such cause can be discovered—where there is no syphilitic history—one must consider the possibility of inherited syphilis, which is capable of producing tabes and general paresis. Heredity in other diseases is not of such great importance, except perhaps in the fact that an unstable nervous system has been transmitted to the offspring, who is more likely to develop disease when exposed to the specific germ, but in syphilis the patient may acquire the infection before birth, and tabes subsequently. No one can deny that fatigue from forced marches and occupations requiring excessive use of the legs may have something to do with hastening the development of tabes. This may explain the greater frequency of the disease among those who are compelled to be on their feet constantly rather than among those who follow more sedentary occupations. Traumatism also may hasten the development of symptoms, but traumatism alone has never been proved to be the cause of the disease, though litigation is common for the recovery of damages for so-called traumatic tabes.

Although tabes occurs most frequently in middle life, there is no period of immunity. It may occur in infants and in men seventy or eighty years of age. Usually an interval of from five to twenty years is required for the development of the disease, but it may appear within half a year or fifty years after the chancre.

Symptomatology.—The symptomatology is so varied and presents such a multiformity of pictures that even a general survey of this disease leads into the general field of neurologic diagnosis. He who masters tabes really knows half of neurology.

The entire course of the disease has been divided into (a) pre-ataxic, (b) ataxic, and (c) paraplegic stages, but in the writer's opinion

such a classification is purely artificial, as there is really no sharp line of demarcation between stages.

COURSE OF A TYPICAL CASE.—We may best begin by describing the usual phenomena of tabes. A patient, otherwise in fair health, may complain of so-called “rheumatic pains” for some months or years before he has occasion to consult a physician. When the pains, which are sharp and cutting, coming and going, and at first depending upon barometric changes, become more severe and intolerable, the patient consults his physician. As the disease progresses, the patient may also notice slight urinary difficulties, that is, he may have to strain in order to expel the bladder contents, or there may be a little dribbling, especially after partaking of alcoholics. A frequent complaint at this time is constipation and perhaps a slight diminution in the sexual powers. Gradually some degree of incoördination may be noticed. The patient perhaps discovers that he is uncertain on his feet in the dark, or when he washes his face in the morning he may notice a slight swaying of the body when the eyes are closed. Somewhat later in many instances, a peculiar sensation of numbness may be complained of in the feet or hands. The ground may feel peculiar, as though the patient were walking on cotton, or as though a soft substance were interposed between his sole and the floor. There may be numbness in the finger tips, especially in the two ulnar fingers. A frequent complaint is a feeling of tightness about the trunk or abdomen, which is often attributed to dyspepsia. These symptoms may last a variable time before the true nature of the disease is recognized. Then there appear the objective signs of tabes, a loss or diminution of the knee reflexes, loss of the Achilles reflexes, Argyll Robertson pupils, anesthetics and analgesias in various parts of the body, Romberg sign, ataxia of station and of motion. In addition, the physician may discover changes in the optic nerve—beginning primary optic atrophy. Rarely there may be the so-called gastric crises, that is, attacks of pain and vomiting lasting a variable time and returning after an interval of freedom from pain. These symptoms may progress rapidly or slowly, becoming more marked in time and eventually terminating in the so-called paraplegic state, when the patient becomes completely ataxic and unable to walk. The sphincter troubles continue, until complete urinary and rectal paralysis occurs. In these later phases the pains may cease, but anesthetics and analgesias with extreme loss of the deep sensibility may take their place.

In exceptional cases optic atrophy occurs early and appears to have a retarding influence on the other classical signs of the disease. At times gastric crises occupy the field without any other symptoms appearing for years. Trophic symptoms in the form of Charcot joint and perforating ulcer of the foot may for a time be the leading symptoms, but in these cases there will invariably also be found many of the other symptoms of tabes.

Having given a brief sketch of the symptoms of an ordinary case of tabes, it will now be of interest to cite, in abbreviated form, a clinical case of that disease which the writer presented before a medical class

in Cook County Hospital, following which the symptoms will be taken up in greater detail.

CASE X.—The patient, 53 years of age, complains of difficulty in walking in the dark and of darting pains which are sharp in character and which come at irregular intervals. There are also peculiar sensations in the hands and feet. They feel numb and dead, as though asleep. The difficulty in walking becomes most noticeable in the dark and when he attempts to walk up and down a flight of stairs. Some time previously he noticed that while washing his face and closing his eyes, he reeled from side to side. He complains of a peculiar sensation of tightness around the chest and of trouble in starting the urinary flow. There is also dimness of vision. As regards sensation, there is paresthesia in hands and feet—a numbness, which, in the legs, is described by the patient as a feeling of cold water running down to the feet. The patient volunteers a history of rheumatism. When questioned as to the character of the rheumatism, he explains that this was mostly localized in the knee and consisted of attacks of sharp pains, which made him jump and “double up.” While he denies gonorrheal infection, he admits a chancre at 25—twenty-eight years ago. He lost weight, a symptom often observed in this disease—the sufferers usually look pale and appear ill. His habits are good; the visceral findings are negative. Objectively, we observe a gait which shows but a slight degree of incoördination when the patient walks across the amphitheater, but in attempting to walk a line or to place one foot in front of the other, he fails completely. Standing with eyes closed and the feet approximated causes him to stagger. This is the so-called Romberg sign.

Thus far the signs and the history point strongly in the direction of tabes. It is now necessary to examine the deep reflexes; they are lost in the knees and in the Achilles tendons. The pupils react sluggishly to light, but normally in accommodation—a beginning Argyll Robertson pupil. The test for sensation brings out the remarkable phenomenon of “delayed conduction of sensation,” that is, he does not feel the pin-prick at first, but after a few seconds he cries out from pain, which he still feels after several minutes. We now test his sense of position and motion. When the patient’s foot is moved passively, he is asked to point his index finger toward his great toe with eyes closed. In this the patient succeeds fairly well on the left side, but not on the right—showing disturbance in the sense of position and motion. In a well-marked case of tabes one often notices a disturbance of the “osteosensibility.” Normally, when a tuning-fork is placed upon a bony part of the body, as, for instance, the tibia, vibrations may be perceived for from 15 to 20 seconds; in tabetics there is either loss or reduction of osteosensibility. This patient does not feel any of the vibrations made with a tuning-fork (small C 128 vibrations); the test is positive. The optic nerve shows a degree of primary atrophy, which explains the visual dimness complained of by the patient. The case is clearly one of tabes.

PHYSICAL FINDINGS.—Now as to the symptoms and signs of tabes. The principal signs are found in (1) loss or diminution of the deep or tendon reflexes, (2) changes in the pupillary light reflexes, (3) disorders in coördination, and (4) in subjective and objective sensory phenomena.

Tendon Reflexes.—Of great diagnostic importance is the finding of a reduction or loss of the patellar or knee reflex and of the Achilles or heel jerk. Of lesser significance is the loss of the triceps or elbow jerk and of the supinator or wrist jerk. In fact, before the arm reflexes show anomalies, other signs of tabes are usually present.

It is mainly with the *deep reflexes* of the *lower extremities* that we are concerned in tabes, for they are almost always present in the healthy and their absence is considered a cardinal sign of tabes.

Knee Reflexes.—The technic is best carried out with the patient in the recumbent posture and completely relaxed. The examiner elevates the knee, allowing the foot to rest flatly on the bed, when he quickly strikes the patellar tendon below the knee; the result is a prompt contraction of the quadriceps extensor muscle. In those unable to relax, Jendrassik's method of reënforcement, or one of its modifications, may be helpful. This method consists in striking the patellar tendon while the patient grasps his own or the examiner's hand, thus putting the upper extremity muscles under tension and relaxing those of the lower extremity. When the unaided method fails, reënforcement often succeeds. In other instances it may be necessary to engage the patient in conversation with an assistant, while the examiner strikes the patellar tendon. A method popular with neurologists, in cases presenting difficulty in eliciting knee jerks, is to have the patient sit on a high chair or table, with the legs overhanging the table and the eyes closed. The everyday method of taking the knee reflexes is to have the patient cross his knees and, requesting him to look straight ahead, the examiner taps the patellar tendons. The normal response is a rapid contraction of the quadriceps, shown by a quick rise and corresponding fall of the leg. If one side is normal, comparison with the opposite side will greatly assist in diagnosis. In tabes the knee reflexes are either pathologically reduced or are absent; in some of the early cases one reflex may be absent, while the other is still present.

The Achilles reflex is important from the circumstance that in tabes it may disappear before the patellar reflex. To obtain the reflex, the patient kneels on a chair, his feet overhanging the anterior edge and the hands grasping the top of the chair, while the examiner strikes the Achilles tendons. The normal response consists in a quick contraction of the posterior leg muscles and an upward movement of the foot. In the recumbent posture the Achilles reflex may be obtained by slightly flexing the knee, turning the leg outward and lightly supporting the toes with the left hand; the examiner then strikes the Achilles tendon with the right. The response is a quick contraction of the calf muscles. Rarely is this reflex absent in normal individuals, and in obese persons it may be obtained with some difficulty.

In tabes the knee and Achilles reflexes are found to be either *reduced* or *absent*.

Pupillary Signs.—Co-equal in importance with the loss of the tendon reflexes is the so-called Argyll Robertson pupil—also called reflex iridoplegia.

Normally when light from any source—diffuse daylight, gas, electricity, or a match—enters the unshaded pupil, there occurs a rapid contraction of the pupil with consequent narrowing of its opening, which varies according to the age and the sensibility of the individual. In tabes the pupil fails to respond to light stimuli, while still accommodating for near vision and in convergence. In examining for this sign one must be careful that patients do not accommodate when light is suddenly thrown into the eye. In order to avoid errors resulting from consensual reaction, it is best to examine each eye separately. The loss of the light reflex may be complete, or there may be only a sluggish response to light—both phenomena being pathological and of equal diagnostic importance.

In tabes the pupils are often unequal and irregular in size, a phenomenon which may for some time precede the development of the Argyll Robertson pupil. Of course, this does not mean that any slight irregularity in the size or shape of the pupil is necessarily an indication of tabes; slight pupillary differences may occur in normal individuals as the result of refractive errors.

Incoördination.—Though tabes has received its popular name—locomotor ataxia—from the fact that ataxia is a prominent symptom, incoördination is not an early sign, but develops gradually and may take a long time for its complete evolution. Romberg first described the symptoms of static ataxia, which consists in the swaying of the patient's body when his eyes are closed and the feet approximated. This is the so-called Romberg sign, a prominent sign of tabes in the somewhat advanced stages. Earlier in the disease the patient himself may have discovered the advent of this symptom by swaying and uncertainty of station, especially when he attempts to wash his face and cover his eyes. In cases of marked ataxia, swaying is noticed when the feet are approximated without closure of the eyes; in those still further advanced, swaying occurs when the patient stands with feet wide apart. The difficulties of maintaining static equilibrium become still greater when the patient stands with bare feet approximated. Then muscular action must replace the support otherwise obtained from the rigid sole of the shoe, and the tendons are seen in constant unrest.

Dynamic ataxia, or incoördination of movement, is occasionally first noticed by the patient when he walks in the dark or attempts to walk backward. When incoördination becomes more accentuated, uncertainty in walking is observed even with the eyes open, especially when the patient attempts to go up or down a flight of stairs. When an ataxic patient is asked to turn quickly, he performs the act very clumsily and may even fall. When incoördination is fully established, the tabetic raises his feet too high, throws them too far forward, brings them down

too suddenly, heel first and toes last, giving the characteristic stamping double-step. As the disease progresses, the patient can walk only with cane or crutch, or is compelled to support himself on another person or an adjacent object. Still later, standing and walking become impossible even with the aid of crutches: the patient becomes bedridden.

In the majority of cases incoördination affects the legs first and foremost; the arms may be involved much later in the course of the disease. Occasionally, however, the upper extremities are first affected; thus the delicate movements used in writing or in the pursuit of skilled labor may show defects.

The usual test for *incoördination* in the upper extremities is to have the patient touch his nose with the index finger while his eyes are closed; first with the right, then with the left hand. When ataxia is present, the attempt goes wide of the mark. Another test for the same purpose is to have the patient, with abducted arms, bring his forefingers together, tip to tip. As the ataxia increases in the hands, patients become less able to properly use their fingers in buttoning their coats, picking up small objects from the table, etc. A test which demonstrates extreme ataxia in the hands is the one in which the patient, with covered eyes, extends his hands with the fingers wide apart, when it is noted that the muscles contract and relax involuntarily, causing a relative change in the position of the fingers. This symptom has been mistaken for athetosis. But no matter how incoördinate the movements are in the extremities, there is never any diminution of power in the muscles themselves. Though there are cases of tabes in which wasting of muscles and paralysis are symptoms, these are complications and not a part of the symptomatology of tabes.

Hypotonia occurs either as a result of ataxia or as an independent phenomenon. The phenomenon consists in an excessive mobility of the joints, owing to reduced muscle tonus. Tabetics have been known to flex the hip on the trunk to an angle of 90 degrees without bending the knee, a feat impossible of accomplishment by an individual not an acrobat. This symptom can best be demonstrated in the abductors of the thigh, when the patient can sit on the floor with the legs almost horizontally extended outward. When hypotonicity of the quadriceps is marked, a patient may be able to touch, with his heels, the posterior portion of his thigh. With involvement of the flexors of the lower extremity there is hyperextension of the knee, a phenomenon frequently observed in tabetics. So characteristic of tabes is the convexity on the back of the tabetic's knee when he is standing that one may almost diagnose the disease from this symptom alone.

Sensory Symptoms.—(a) *Subjective Sensory Findings.*—One of the most troublesome symptoms of tabes is the appearance, at irregular intervals, of sharp, cutting pains. These attack one portion of the body after another, may be of momentary duration and leave behind them a feeling of tenderness and soreness of the skin. Because the pains are frequently aggravated by low barometric pressures, they have often been considered rheumatic in character, both by patient and physician.

While in some cases the lightning-like pains are so mild that the patient does not even mention them, there are cases in which these pains constitute the principal complaint. They are shifting in character, often described as millions of needles and pins, starting below the knee and radiating into the toes.

In addition to these pains there are present sensory symptoms described as *paresthesia*. These are peculiar sensations of formication, tingling, or numbness, which may appear early in the disease and constitute a most annoying symptom. The feeling in the feet is often described by the patient as of cotton being interposed between his skin and the floor. The paresthesia may appear spontaneously or after slight irritation, as when the nerves of the foot are pressed. In the upper extremities a favorite location for paresthesia is in the ulnar nerve distribution of forearms and hands, which feeling the patient describes as numb. In the trunk the sensation is mentioned as a tightness around chest or abdomen, the so-called "girdle sensation," which may appear after a full meal or independent of the ingestion of food.

(b) *Objective Sensory Findings*.—Subjective sensory symptoms are usually accompanied by those signs which can be determined by the physician's objective examination, and which are therefore called objective sensory findings, though in reality all sensation is subjective because dependent upon the patient's statement or gesture.

Among these sensory disturbances *analgesia* and *anesthesia* of the lower extremities, as well as *hypesthesia* over the trunk, are most frequently encountered. Because of its importance, it is well to briefly discuss the methods for testing analgesia. The simplest method is to ask the patient, previously divested of his clothing, to indicate when he feels the examiner's pin-prick applied lightly to the skin. The normal response is immediate and should be compared with that given for a corresponding spot on the opposite side. Statements as to sensory findings become of value only after repeated tests have been made with practically identical results. In this connection the reader must be reminded of the fact that some few normal individuals are insensitive to various stimuli, but as a rule analgesia is a pathological phenomenon and found in a large number of tabetics. So profound may the analgesia become that a pin introduced deeply into the skin may be perceived by the patient as mere pressure or touch. And not only the skin, but also the parts beneath the skin may become analgesic. Indeed, deep analgesia is almost a characteristic sign of tabes, while superficial analgesia may be only indicated or entirely absent. Analgesia of the deep parts may be sufficiently intense to permit fractures of bones and deep skin ulcerations to occur without the patient experiencing any pain whatever. Analgesia, though most frequent in the lower extremities, is also encountered in the trunk, especially in the vicinity of the nipples. Though loss of the pain sense—analgesia—is more marked than anesthesia, or loss of the tactile sense, the reverse obtains in the trunk, where touch is usually more disturbed than pain. Instead of complete loss, there may be only reduction, so-called *hypesthesia* or *hypalgesia*, or there

may be present "delayed conduction of sensation," that is, a pin-prick is first felt as a touch and later as pain. The symptom of insensitiveness of the testicles, as well as analgesia of the various nerves, is only one part of the numerous sensory disturbances occurring in tabes. Analgesia of the ulnar nerve, first described by Biernacki, and since called *Biernacki's sign*, is another of the valuable signs of tabes. It is easily elicited by placing the finger between the internal condyle of the humerus and the olecranon process of the ulna and applying light pressure. While this is keenly felt by a normal person, the tabetic experiences little or no pain. Analgesia of the nerves supplying the bladder may explain the urinary difficulties of tabetics. It is well known that in the early stages of tabes patients hold their urine too long—appear to have no desire to empty the bladder, there being a relative degree of retention—while in the more advanced stages there is complete analgesia with consequent incontinence and dribbling of urine. Disturbances similar in character are observed in the rectal sphincter; but here obstinate constipation is the rule. Occasionally sphincter difficulties may be detected after administering cathartics when calls to the toilet become premature, or too late to be of use, there being involuntary losses of the bowel contents. The sexual functions also suffer from general analgesia; sexual desire is lost early in the disease.

Visceral Crises.—Among the stormiest and most disagreeable sensory phenomena of tabes must be reckoned the so-called crises. Because of their frequency and severity a short description must be given of the gastric and laryngeal crises, though clitoris, rectal and anal crises, as well as liver, urethral and kidney crises are troublesome but rare.

Gastric Crises.—These are attacks of nausea and vomiting accompanied by intense pain, coming on at irregular intervals and causing extreme prostration. During their continuance the patient is unable to take nourishment, the sight or smell of food may bring on fresh attacks of vomiting. The interval between attacks may vary from days to years and is not generally conditioned by indiscretions in diet. During a crisis there is agonizing pain, beginning in the epigastric region, which may extend to back, shoulders, sacrum and chest. First there is vomiting of stomach contents, later bile-tinged material appears, and toward the end of the attack there is prolonged retching unaccompanied by vomitus. A single attack may last from a few hours to as many weeks, at the end of which time the patient is completely exhausted and appears ready for the grave. Almost as suddenly as it comes does the crisis disappear. The pains, nausea and vomiting cease and a ravenous appetite develops. Within a few days, or at most a week or two, the patient regains his strength and resumes his occupation. After an indefinite interval of time another attack appears as suddenly as the previous one and just as suddenly disappears.

Not all gastric crises are accompanied by vomiting and pain. There may be attacks of violent pain without vomiting, which, as regards severity, are comparable to the attacks caused by ulcer of the stomach;

and, on the other hand, there are attacks of vomiting unaccompanied by pain.

The periodic and sudden appearance and disappearance of stomach pain and vomiting is of great diagnostic significance and should create a suspicion of tabes, even in the absence of other signs of that disease. Indeed, it may appear as the earliest symptom of tabes and before any of the others. For years some of these patients have consulted numerous specialists; some of whom have advised and others have actually performed operations for supposed gall-stones and even gastrostomies have been made on these unfortunates. In this connection it is instructive to consult Nuzum's interesting article on "Needless Surgical Operations from Failure to Recognize Tabes Dorsalis." Happily for the patients, surgeons are learning to examine for tabes and mistakes are constantly becoming fewer.

Laryngeal Crises.—Only second in importance to gastric crises are the laryngeal crises. Like the gastric crises, they have a sudden beginning and an abrupt ending. Attacks vary in severity; there may be merely irritation in the larynx with spasmodic cough of variable duration, or there may be inspiratory dyspnea similar to that observed in pertussis. In the more severe cases suffocation, loss of consciousness and generalized convulsions appear. An attack of laryngeal crisis presents a picture even more alarming than the most violent gastric crisis.

Optic Nerve Atrophy.—In about 10 per cent. of tabetics the optic nerve is affected by primary atrophy terminating in blindness. The optic nerve appears pale and vision is gradually lessened. As in the sphere of general sensation, subjective symptoms precede the objective signs of visual disorder. Patients complain of cloudy vision and of a feeling as though a fog or a veil obscured their sight. Sooner or later, however, pronounced defects appear in the visual fields, or the color fields show losses, at first for green, then red, and lastly blue. Vision may be preserved as long as the temporal fibers remain intact, though the ophthalmoscope may already reveal a pale disk. The course of optic nerve atrophy is toward complete blindness, but remissions occasionally occur lasting years. In some cases blindness occurs rapidly. Both eyes are usually affected simultaneously; in some instances one eye becomes blind before the other. In cases of pronounced optic atrophy the ophthalmoscope reveals a grayish discoloration of the papilla and narrowing of the blood-vessels, while the lamina cribrosa appears more prominent than in the normal eye. The entire process may be limited to the temporal half of the optic nerve. A remarkable clinical fact has been noted in connection with the development of optic nerve atrophy in tabes, namely, that if the atrophy is the first symptom to appear, other symptoms of tabes are either late in coming or are very few in number. In advanced cases of tabes, optic nerve atrophy is seldom encountered.

Ocular Palsies.—In the beginning of tabes, ocular palsies may appear as transient phenomena. For a short time diplopia or double vision, combined with or without ptosis, may be noticed. A single muscle or an entire group of muscles may be involved, and external ophthalmo-

plegia may dominate the picture. Most often the entire oculomotor or third nerve is selected by the paralytic process, though in many instances the internal eye muscles alone suffer the brunt of the attack; then we have dilated pupils and loss of accommodation. Transient unilateral or bilateral ptosis may also occur, which may disappear almost as quickly as it came. There may not be a complete paralysis of the eye muscles, but slight weakness of the levator palpebra may become manifest by a lagging behind of the upper lid after exercise. In other instances the weakness of the eye muscles cannot be demonstrated objectively, but the patient complains of subjective fatigue in them.

Cranial Nerve Palsies.—Paralysis of other cranial nerves is not common, with the exception of the trigeminus and the vago-accessorius.

Trigeminus involvement is manifested by anesthesia of the skin of the face, as well as of the adjoining mucous membranes of the nose and mouth.

Paralysis of the vago-accessorius, if limited to one side, is not difficult of recognition; the muscles affected are the laryngeal dilators and constrictors—more frequently the latter, which do not produce threatening symptoms. Indeed, the milder grades of unilateral laryngeal paralysis may occur without symptoms, being often an accidental finding in a systematic examination for tabes. It is the bilateral variety of paralysis of the glottis dilators which causes alarming symptoms in the form of stridor and dyspnea, produced by the slightest exertion on the part of the patient. An unfavorable feature of these palsies is that, unlike those of the oculomotor group, they show no tendency to recede, but become permanent.

Peripheral Nerve Disease in Upper and Lower Extremities.—This is a rare finding in tabes. The reason for its occasional appearance is that, because of anesthesia and loss of the pain sense, patients are less able to guard against trauma than normal individuals. In the lower extremities the peroneal group of muscles is most frequently affected, while in the upper extremities the paralysis is found in the radial, median and ulnar territories.

General Weakness.—Tabetics often suffer from a form of general weakness, not amounting to paralysis, which may either be the expression of general debility or the result of ataxia.

Trophic Disturbances.—Disturbances of general nutrition, anemia and emaciation, even pronounced marasmus, atrophy of muscles, with tendency to bed-sores, are observed in tabes of long duration. Rapid loss of weight is not uncommon in any stage of the disease.

Of greater importance than the general weakness of tabes are the local trophic disturbances, notably so-called *perforating ulcer* of the foot, *pathological fractures*, and *arthropathies*.

In the majority of cases *perforating ulcer* occurs in the foot, rarely in other parts of the body. The ulcer is usually circular in outline and has a tendency to perforate the skin and subcutaneous tissues down to the bone, causing ulcerations which are slow to heal. In the depth of the ulcer are seen pale granulations, from which exude purulent secre-

tions. Its common location is the plantar surface of the foot, especially the base of the big toe, though it may be found anywhere on the plantar surface of the metatarsal region. In still other cases there is a resemblance to a corn which has gone on to suppuration. Sequestra may be thrown out from the depth of the ulcer when the bone has become implicated in the process.

Spontaneous fracture is an uncommon but characteristic symptom of tabes. The word spontaneous as applied to these fractures is really a misnomer, as these fractures never occur spontaneously, but as the result of causes inadequate to produce fracture of a perfectly healthy bone. Such fractures usually heal with an abundance of callus formation about the fractured ends, and little or no pain is ever experienced by these patients at the seat of fracture, though in a few isolated cases pain may be complained of. Nearly always one bone is affected at one time; very rarely is there involvement of more than one bone.

With spontaneous fractures there may be seen other trophic changes in the joints, especially in the knee joint; thus, arthropathy and pathological fracture may occur together early in the disease.

Arthropathy may begin with an acute serous exudate which may be slowly absorbed or may remain; and when the fluid is artificially removed there is a tendency for rapid recurrence. As the processes continue, superficial destruction of the ends of the joints occurs, causing fragments to loosen, which give a peculiar grating and crepitus to the hand when applied to the joint in motion. The roughness of the joint ends is increased by bony proliferation, causing first a hypertrophy and later, when the bone becomes absorbed, atrophy. Owing to thickening of the joint ends and consequent luxations, peculiar grotesque deformities of the joints may appear. The parts in the neighborhood of joints may also become implicated and great swelling develop. All kinds of deformities may be discovered at the knee joint—genu valgus, varus and combinations. Loose portions of the joint may become consolidated by ankylosis, and a certain degree of usefulness may thus be given an otherwise useless joint. Arthropathy and spontaneous fracture may occur together.

VARIOUS STAGES OF THE DISEASE.—In the majority of cases the course and progress of the disease are rather slow, and several periods may be distinguished clinically. There is (1) the so-called *pre-ataxic* period, during which there is no incoördination; (2) the *ataxic* period, in which standing and walking become more difficult—the patient becomes ataxic; and (3) the period when standing and walking become impossible, and the patient becomes *paraplegic*. As previously remarked, there is no sharp line of demarcation between these stages; they are merely artificial creations for purposes of clinical description.

(1) *Pre-ataxic Period*.—At this time the patient usually suffers from so-called lightning pains, interpreted by the sufferer as rheumatic, for which he may not even seek medical advice. When these pains reach a certain degree of intensity, a physician may be consulted who often confirms the patient's diagnosis and prescribes for rheumatism. In some

cases other symptoms may attract the physician's attention to the nervous system as the possible seat of disease. He may discover a slight ocular palsy, slight diminution of vision, some incoördination, or a gastric crisis. The patient himself may notice some uncertainty in the dark when he seeks the toilet-room at night, or he may experience a feeling of insecurity in turning about quickly. There are cases in which the symptoms are mild, indefinite, few and stationary; this is the benign type of the disease. In by far the larger number of cases, after three to five years from the beginning of the disease, the symptoms become aggravated, more ataxia develops, and the patient enters the second or ataxic stage. In a small number of cases the second stage appears more abruptly—the patient develops so-called acute ataxia.

(2) *Period of Ataxia*.—At this period of the disease incoördination of movement in the lower extremities is more marked, the ataxia becoming gradually worse and subsequently invading the upper part of the body. Visceral symptoms and sphincter disturbances become more noticeable and the symptoms may remain more or less unchanged for years, or may gradually merge into the paralytic stage. In some few instances the ataxic period comes to a close suddenly, ushering the patient rapidly into the third stage.

(3) *Paraplegic Stage*.—In this period the patient loses control over the movements of the lower extremities. He is obliged to remain in bed or in an invalid chair, and becomes totally powerless. There is also complete loss of sphincter control, urinary, rectal, and sexual. Besides, the trophic changes in the skin become troublesome; there may be deep ulcerations over the buttocks and numerous bed-sores develop from insignificant pressure over the bony parts. Strangely enough, patients may live many years after they have entered the paralytic stage. This is not the rule, however; in the majority of cases patients in this period show lessened resistance to all sorts of infections, pneumonia, cystitis, and septic poisoning. The termination is usually by acute infection or sepsis.

In discussing the various stages of tabes we must remember that a natural arrest appears to take place in numerous cases; a patient may suffer excruciating pains for many years without becoming ataxic; and ataxic patients may be seen going about the streets for an indefinite time. Many of the symptoms, such as ocular and other palsies, may disappear, especially early in the disease. The palsies, occurring late in the disease, usually remain more or less permanent. Ataxia, when thoroughly established, rarely disappears, though improvement is possible after adequate treatment. The deep reflexes when once lost never reappear; the Argyll Robertson pupil also remains as a perpetual sign of the disease.

LABORATORY FINDINGS.—It is quite possible for a patient to suffer vague pains which go under the name rheumatic, or from symptoms designated as neurasthenic, until the laboratory makes a correct diagnosis by reporting a positive Wassermann on the spinal fluid or blood, or both.

According to Kaplan, the spinal fluid changes are almost as constant

in tabes as the Argyll Robertson pupil or the absent knee jerks. The laboratory is frequently able to furnish reliable evidence when the clinical diagnosis is most obscure, but the rule is that in diagnosis the laboratory usually plays an insignificant rôle. Its greatest usefulness lies in the gauging of therapeutic progress. Many times it will also be found that the blood Wassermann is positive, while the spinal fluid is negative. On the other hand, with Nonne and Hauptmann's method of testing the spinal fluid, the number of positive fluids should become considerable. In addition there is an increase of globulin and of lymphocytes—lymphocytosis.

There is a laboratory type of tabes which Kaplan has called the "*hyperlymphocytic type*," in which the cell-count in the cerebrospinal fluid is high. It is generally stated that the higher the cell-count, the more active the disease and the more violent the symptoms. In the majority of cases of tabes the spinal fluid yields a positive Wassermann with the larger amounts of fluid, say from 0.8 to 1 c.c. (Hauptmann's method), and also shows a lymphocytosis. With a high cell-count one usually encounters an excess of globulin.

Another laboratory type of tabes has been described by Kaplan as the "*negative type*." In this there is an absolutely normal serology with an insignificant pleocytosis (lymphocytosis) from 12 to 32 cells per cu.mm. The "negative type" serology is indicative of the absence of meningeal irritation and is a sure sign of a more or less purely degenerative process. While this is the rule, there are exceptions; a "negative fluid" tabes may show clinical evidences of active inflammatory processes.

Again, according to Kaplan, there is a "relatively negative type" of tabes in which there may be a moderate cell-count, say 25 cells per cu.mm., while the remainder of the serology is negative. Another variety of serology shows a negative Wassermann reaction in the blood, a positive reaction in the cerebrospinal fluid, a normal globulin reaction, and a pleocytosis of from 20 to 50 lymphocytes per cu.mm. This variety is observed after treatment.

Still another laboratory type is "*Wassermann fast*" tabes, which means, a type of tabes showing a great resistance to treatment, at least as far as laboratory improvement is concerned. Especially has Kaplan noted in these cases that the blood-serum does not change from a positive Wassermann, while the spinal fluid frequently becomes negative. The question might well be asked, as to whether this type is one which will **gradually merge into** general paresis. The answer will be given when our experience becomes larger and when every case of neurosyphilis is subjected to a thorough serological examination before beginning treatment.

Diagnosis.—Tabes may be easily diagnosed when symptoms are in abundance, but it may be one of the most difficult diseases to diagnose when symptoms are few and ill-defined. The greatest stress must be laid on an examination of the pupils, which gives perhaps the most valuable information. One observes their size and whether they are equal

and regular and if the light reflex is normal. It must not be forgotten that long before this reflex is lost, there will be found a sluggish response to light stimuli, or the light reflex may be lost in one eye only. The Argyll Robertson pupil is an important diagnostic sign of tabes. Of course, the absence of the accommodation and light reflex does not militate against tabes, though the so-called "fixed" pupil is more characteristic of the earlier stages of neurosyphilis.

Perhaps next in importance is the so-called Westphal sign, or the absence of the patellar reflexes. There are all grades of pathological knee reflexes; there may be only a reduction or a loss of one reflex, a finding which has diagnostic value, unless produced by a local nerve lesion. The rule in tabes is bilateral loss of knee jerks, while in other forms of neurosyphilis we are more apt to find inequalities. Of equal, if not of greater, importance is the early loss or reduction of the Achilles reflex. To be of value in the diagnosis of tabes this loss must be bilateral, for unilateral loss of the Achilles reflex is a common sign of sciatic neuritis. The third great symptom of tabes is reduction or loss of co-ordination. This is first noted in the sign of Romberg, that is, when the patient stands with eyes closed and heels and toes approximated, he sways considerably; this is in marked contrast to the normal individual whose swaying is minimal. Of course, we do not at this time consider that form of advanced ataxia in which the patient can only walk with the support of canes, eyes fixed on the floor and body bent forward, gait stamping, with the double step of heel first and toes later. Even a layman is able to recognize tabes when the gait has assumed such characteristics. There are cases, however, when the ataxia is incipient and unknown to the patient himself. There may be some difficulty in the finer movements of both lower and upper extremities, which only the physician can bring out by the proper tests. Of course the finding of ataxia, if polyneuritis and cerebellar disease can be excluded, is of extreme diagnostic significance for tabes.

Primary progressive optic atrophy is caused by but few other conditions besides tabes, and its determination is almost equivalent to a diagnosis of tabes.

In discussing the characteristics of the subjective sensory disorders of tabes it was sufficiently emphasized that the lightning quality and the intermittency, with the intensity of the pain, all have diagnostic value for tabes, though root irritations from other causes may produce similar pictures. The objective sensory disorders, with the special predilection for the deep sensibility, and the peculiar segmental distribution, both as regards analgesic and anesthetic disturbances, are in themselves suggestive of tabes, though other conditions cannot be excluded.

In the diagnosis of tabes one always thinks of the cardinal signs of the disease: (1) loss of tendon reflexes, (2) pupillary changes, (3) ataxia. If all three symptoms are absent, tabes is improbable. Whenever a patient suffers from severe, lancinating pains coming on at irregular intervals, suspicion of tabes should lead to search for one or more of the cardinal signs. On the other hand, if in a routine examination of

a patient one should accidentally discover *one* of the cardinal signs, a diligent inquiry should be made for other symptoms of tabes. Efforts in this direction will seldom be in vain; there will usually be found a history of transient ocular palsies, or objective sensory disturbances—perhaps Biernacki's sign, trunk anesthesia, or hyperesthesia to cold, may be ascertained. There are all sorts of combinations of symptoms—the so-called cardinal signs and a host of secondary symptoms. For instance, a patient may suffer from the lightning-like cutting pains and show inequality and irregularity of the pupils besides, and on inquiry he may reveal a definite history of syphilis. Such findings will no doubt suggest further steps in the diagnostic inquiry. A lumbar puncture is made and the spinal fluid examined for Wassermann, globulin increase, and lymphocytosis. As has been repeatedly emphasized, a negative laboratory finding by no means excludes the disease, while the positive finding makes it extremely probable.

In the order of diagnostic importance the symptoms may be thus enumerated:

- (1) Loss of the tendon reflexes.
- (2) Argyll Robertson pupils, complete or incomplete.
- (3) Sensory disturbances (lancinating pains and objective findings, anesthetics and analgesias).
- (4) Romberg sign.
- (5) Ataxia of locomotion.
- (6) Hypotonicity of muscles.
- (7) Sphincter disturbances.
- (8) Paralysis of ocular muscles.
- (9) Optic atrophy.
- (10) Crises (gastric and laryngeal principally).
- (11) Trophic changes.

DIFFERENTIAL DIAGNOSIS.—The new laboratory tests for neurosyphilis, generally believed to aid in diagnosis, do not always solve the difficult problems of differential diagnosis. It is true that when in doubt as to the specific origin of a certain neurologic lesion the blood and spinal fluid examination may give valuable decisions. Even in this respect the laboratory may decline the responsibility of making a diagnosis, by simply furnishing a negative finding. Certainly the laboratory does not undertake to differentiate the various types of neurosyphilis except in a very modest way, as outlined under Laboratory Findings (pages 109-110).

Take, for instance, syphilitic *meningomyelitis*. When the process affects the posterior portion of the cord first or principally, the symptoms may be identical with those of tabes. Clinically we may have sensory symptoms, ataxia, and in certain instances even eye findings much like those found in tabes. Careful scrutiny will disclose marked differences. First, as to the sensory findings, the pains will be more continuous and limited to root territories and there are no gastric crises in meningomyelitis, while in tabes the pains are of the lightning-like

variety and are exceedingly transient; further, gastric crises may occur. Ataxia, if it occurs at all in meningomyelitis, is slight and does not present the well-known Romberg sign and the dynamic coördination disorders of tabes. As regards eye findings, they will be those of active syphilis, namely, "fixed" pupils; only cervical or upper dorsal syphilitic meningomyelitis may yield the Argyll Robertson phenomenon, but in this localization there will also be spasticity and exaggerated reflexes—findings foreign to tabes. On the other hand, the lumbar form of meningomyelitis will yield absent reflexes and also muscular atrophies—the last of which are not seen in tabes. Naturally all the symptoms of this active form of the interstitial type of syphilis are acute in their onset, and serious doubts may be dissipated by a little patient waiting. Serologically the percentage of positive Wassermanns is higher in meningomyelitis than in tabes, and lymphocytosis in the spinal fluid reaches its highest intensity in the active cord lesion, while in tabes the lymphocytosis is only moderately high.

Spinal cord tumor may occasionally simulate tabes, especially when the growth begins in the posterior half of the cord. The differential diagnosis will be made when a distinct level of anesthesia is discovered and when absence of all symptoms, motor, sensory, and reflex, is noted above the same level. This certainly excludes tabes, in which signs and symptoms are scattered over a wide area. Then again, a spinal fluid examination will yield invaluable evidence in differentiation between a non-specific tumor and tabes.

Syringomyelia may rarely give rise to difficulties in differential diagnosis when the lumbar cord is affected. Here the pupillary signs of tabes are absent and the sensory dissociation of syringomyelia rarely or never occurs in the syphilitic cord disease. Serum or spinal fluid Wassermann has never been seen in syringomyelia.

Subacute combined cord degeneration may produce a picture resembling tabes, especially when the spastic symptoms from pyramidal tract involvement are over-shadowed by those of posterior column degeneration. It will be found, however, that even at an early stage of the disease there will be added to the ataxia exaggerated and pathological tendon reflexes, such as Babinski and Gordon signs. Besides, Argyll Robertson pupils do not constitute a part of the symptomatology of combined cord disease, while they are almost always found in tabes. In addition, the characteristic blood-picture of pernicious anemia, which is a frequent accompaniment of subacute combined cord disease, is not seen in tabes. Of course, the syndrome of ataxic paraplegia may be produced by syphilitic cord disease of the interstitial variety; then the criteria given for differentiation of the interstitial from the parenchymatous form of syphilitic cord disease may be applied.

Friedreich's ataxia may be confounded with the form of tabes occurring in young individuals as the result of hereditary syphilis, the so-called *juvenile tabes*. In Friedreich's ataxia there is the family and hereditary element, absent in tabes, the presence of normal pupils and the absence of subjective and objective sensory disorders, including

sphincter losses—all of which are different in tabes. In addition, the lower extremities are more often involved in tabes, while the upper extremities are early affected in hereditary ataxia. The latter disease almost always shows evidences of pyramidal tract involvement in addition to cerebellar ataxia, while the ataxia of tabes is distinct from cerebellar ataxia and is unaccompanied by Babinski or other pyramidal tract signs.

In *cerebellar tumor* we occasionally encounter symptoms resembling tabes. For instance, there may be loss or reduction of the deep reflexes and ataxia. However, in tumor we shall probably also find vertigo, vomiting, optic neuritis, and the absence of Argyll Robertson pupil. The ataxia of cerebellar disease is different from tabes in that the former consists more of a side-to-side swaying, while tabetic incoördination, for the most part, presents an uncertainty in the anteroposterior direction. Again, the laboratory findings of tumor are different from those of tabes.

Perhaps the greatest difficulties in differential diagnosis arise between true tabes and so-called *pseudotabes*, a form of multiple neuritis with tabetic symptoms. Ataxia is found in both and also absence of deep reflexes; the sensory disturbances also resemble each other. Indeed, we may even see sluggish pupils in multiple neuritis, but these are not permanent—disappearing as a rule after the toxins have been removed from the body—and rarely are they of the Argyll Robertson type. In polyneuritis the pains are not lancinating and shooting, but usually they are dull and more or less continuous; there are no girdle sensations in multiple neuritis. The muscles in multiple neuritis may show ataxia, but there will invariably be found evidences of paralysis or paresis in the form of disability and wasting of the lower extremities. Then the finding of an etiological element in the form of alcohol or acute infectious disease, such as diphtheria, or the presence of diabetes, will aid materially in differentiation; and last, but not least, the blood and spinal fluid examinations will settle the diagnosis.

Clinical Varieties.—The usual types of tabes having been described, there remain to be mentioned certain varieties of tabes which are rarely seen, but must be recognized.

First, there is the *bulbar type*, in which cranial nerves are involved early. The subject of this variety of tabes may be a sufferer from laryngeal or pharyngeal crises coming on at irregular intervals. Somewhat later ocular palsies and optic atrophy make their appearance. Indeed, optic atrophy may for a long time be the only symptom. Sooner or later other symptoms may become associated with the optic atrophy, but rarely do cord symptoms appear, and certainly ataxia is exceptional in this type of tabes. There are cases with trigeminus neuralgia, that is, those in which the sharp shooting pains occur in the face. In these cases also anesthesia and analgesia seem to be limited to the facial territory.

Then there is a so-called *cervical type* of tabes in which the disease affects principally the upper part of the body: there is extreme ataxia

in the arms and there are pains radiating into the territory supplied by the segments of the cervical cord. In these cases also the triceps and supinator reflexes are absent, while the deep tendon reflexes of the lower extremities may still be present. In addition there are present the usual ocular phenomena of tabes, such as Argyll Robertson pupil and perhaps diplopia or ptosis, but the sphincters may be found intact. Another type of the disease is represented by *superior tabes*, in which there are both bulbar phenomena and cervical cord symptoms. A type which is rare and consequently seldom recognized is the *conus* and caudal type of tabes. In this last variety the symptoms are confined to the small territory supplied by the conus proper; we may thus have limited anesthesia in the region of the perineum, sphincter disturbances, trophic changes, etc., but no symptoms from dorsal or lumbar cord involvement.

The variety described as *juvenile tabes* is tabes occurring in the young. The patients are between 6 and 25 years of age and the offspring of syphilitic parents. The symptomatology does not differ materially from that of the acquired form of tabes in the adult. It has been noted, however, that many cases begin with urinary troubles, some with lightning-pains, and few with blindness due to optic atrophy. Though gastric and intestinal crises are relatively common in this variety of tabes, ataxia is usually slight and tardy in appearing, while marked ataxia is exceedingly rare.

To further elucidate some of the clinical varieties of tabes the writer will incorporate here the histories of two clinical cases, one a case of superior tabes, the other one of juvenile tabes.

CASE XI.—*Case of Superior Tabes*: The patient, a married peddler, entered Cook County Hospital for the relief of pains. His health history was negative as to visceral disease, but he admits a syphilitic infection in 1888, for which he was treated with mercury during an entire year. Habits were irregular: until recently he smoked and drank alcoholics excessively. The beginning of the present trouble is dated six years ago, when he developed sharp, shooting pains, at first under the right scapula, later under the left shoulder blade. The pains came in bouts, were cutting and of momentary duration with irregular intermissions; each attack consisted of a series of lancinating pains with irregular pauses between them, and lasted from hours to days. In addition, there were spells of vomiting accompanied by excruciating pains, corresponding to what is known as gastric crises. While up to a few months ago there were periods of complete freedom from attacks, the latter have become more frequent and more severe. The pains now radiate in all directions, upward and downward, affecting arm and fingers, as well as legs and toes. During the last few months the patient also experienced pain in the left half of the face in front of the ear, which pains shoot downward from the vertex of the head to the point of the chin. There are similar attacks of pain in the epigastric region, lasting a few hours, and accompanied by vomiting. The patient is

constipated and suffers from urinary difficulties, retention and incontinence. About two years ago ocular palsies were noted in the left eye and, at the same time, failure of vision became apparent. With the eye symptoms there appeared certain paresthesias described as peculiar sensations of heaviness and numbness in the left half of the face; there was also some weakness of the muscles of mastication on the same side.

Examination.—The patient is fairly well-nourished; the viscera are normal. A left-sided ocular ptosis and left external rectus paralysis are in evidence. The pupils, rather small and slightly irregular in outline, are of the Argyll Robertson type. There is a bilateral beginning optic atrophy. The teeth are absent on the left side, the patient having lost them about two years ago. He states that when his left molars began to loosen he had no difficulty in extracting them with the fingers; that the upper teeth had also become loosened, but eventually tightened and now appear crowded upon each other. The entire process was painless. His hearing is reduced in the left ear.

Reflexes.—The superficial reflexes are easily elicited. The deep reflexes appear normal in the upper extremities. The knee-jerks are brisk, the left slightly in excess of the right; Achilles tendon gives good response bilaterally. Masseter jerk is present, but the McCarthy reflex is absent on the left side and also the left scapulohumeral reflex.

Coördination.—There is no incoördination in the upper extremities. Romberg's sign is absent; there is no distinct ataxia of movement, except a slight degree, which becomes noticeable when the patient hops on one foot.

Sensation.—There is a slight reduction of tactile sensation on the left side at about the level of the nipple, extending circularly around half of the body. There are present left ulnar analgesia and right ulnar hypalgesia. Reduction of the pain sense is present in the left half of the face: pin pricks are not perceived as keenly as on the opposite side. In addition to the subjective feeling of numbness and heaviness in the left half of the face there is hypersensitiveness to cold, but heat can be differentiated from cold. There are no sensory anomalies in the buccal mucous membrane; and taste is not impaired. Muscle and joint sense in both upper and lower extremities seem perfectly normal.

Summary.—Lancinating pains, Argyll Robertson pupils, primary progressive optic atrophy, paresthesia in face, falling out of teeth, ocular palsies, bladder disturbances—constitute a group of symptoms sufficient to establish the diagnosis of tabes. Interesting is the fact that almost all the spinal deep reflexes were preserved, and coördination was practically unimpaired. The symptoms making the diagnosis certain were mostly found in the head and face. The case is typical of what is called "superior tabes."

In order to acquaint the reader with the picture of what the writer considers a typical case of *juvenile tabes*, the history, in part, of a case which the writer reported in the *Journal of Nervous and Mental Diseases*, 1904, is here presented:

CASE XII.—Case of Juvenile Tabes: The patient, 25 years of age, single, was a telegraph operator until about four years before this examination.

Family History.—Father died of some lung trouble at 47; mother, of tabes at 57; one brother, of juvenile general paresis at 20; one sister, of interstitial neurosyphilis, gummatous meningitis and hemiplegia at 29.

Health History.—Patient was born at full term in normal labor. When about two or three months old, he is said to have had moist papules around the anal margin and rhagades about the oral ring. At the age of four months he had a generalized eruption. He had measles at about six years, and typhoid fever eight years ago. At fifteen he had what he calls "shingles," a vesicular eruption. When about five years old, and up to his tenth year, he suffered from enuresis nocturna and diurna. After this there developed difficulty in emptying the urinary bladder, slight at first, but more aggravated later.

Habits.—For two years, up to quite recently, he was a heavy drinker. For the past four months he has been a total abstainer from alcoholics. He has never sustained any injuries.

Present Illness.—In 1896, he accidentally discovered that his right eye was totally blind. Shortly thereafter vision began to fail in the left eye, and during the last year and a half he has been completely amaurotic. About five years ago he developed characteristic lightning-like pains in the lower extremities. They were very severe at first, and each attack would last from two to three days, with slight remissions. At the present time he is only infrequently annoyed by these pains, and they have lost their former severity. A peculiar numbness and tingling in the lower extremities appeared soon after the shooting pains had begun. Added to this there was a feeling as though he were walking on rubber. Quite recently he experienced pains which began in the hypochondriac region and extended almost circularly to the spine, and which gave him the sensation of a band tightening about his body.

Status Præsens.—General appearance rather youthful, beardless and feminine. No asymmetry of face and skull. Radiating from each corner of his mouth are the silvery white lines typical of inherited syphilis. The teeth are well formed and there is an abundance of pubic hair, but the testicles are infantile in size. Heart and lungs are normal; the pulse is regular. The abdominal viscera are normal. Bony deformities and peripheral arteriosclerosis are absent; the superficial inguinal glands are slightly enlarged, but no epitrochlear or cervical glands can be felt. There are no visible scars on the trunk, but at the anal margin are distinct radiating lines, probably scars of former condylomata. There is a perforation of the nasal septum.

Eyes.—As long ago as 1900 there was complete optic atrophy in both eyes. Vision of the right eye, hand movements; of the left, 5–200. Fields of vision showed marked contraction for white, with doubtful color perception.

At the present time there is a slight dropping of the left upper eye-

lid and paresis of the left internal rectus; the eye does not rotate beyond the middle line when the patient looks to the right. When this oculo-motor paresis first occurred is unknown—but evidently after he became completely blind, or diplopia would have been noticed. Both pupils are widely dilated, the left a trifle more so than the right; neither responds to light or to accommodation. The blindness is so complete that not even light can be distinguished from darkness. Both optic nerves present a snowy white appearance with distinct definition, and almost total disappearance of the vessels.

Muscular Weakness.—With the exception of the above-mentioned partial paresis of the motor oculi, no muscular weakness can be detected; and there is no tremor in fingers and hands.

Sensation.—Slight paresthesia, lightning pains and girdle sensation are complained of.

Objective.—Tactile: pain and temperature senses are intact, except that in a few isolated spots on the anterior surface of the legs a pin prick is first noticed as a touch, and a little later as an intense and prolonged pain. Firm pressure of the right testicle causes a sickening pain; pressure of the left is first felt as touch; a little later the patient cries out from pain. The sense of position is unimpaired in the hands; in the toes of the right foot flexion is mistaken for extension; and he is entirely unable to indicate the position of his left toes. Osteo-sensibility is greatly reduced, according to Egger's tuning-fork test.

Coördination.—Ataxia of station and of motion, already noticed by the ophthalmologist in 1899, is well marked. Though not extreme, it can be readily elicited by the heel-to-knee test. In the upper extremities the ataxia is less marked on the right than on the left side.

Under this heading might also be mentioned a peculiar ataxia of the tongue and facial muscles. When the patient protrudes the tongue there is a constant forward and backward swaying of the tongue, resembling somewhat the lateral swing of a pendulum. It is coarse, and dissimilar to any tremors previously seen by the writer.

The facial muscles show a peculiar quiver when in action; this is an ataxia of facial and tongue muscles.

Reflexes.—Abdominal and plantar reflexes are normal; the cremaster cannot be obtained on either side. McCarthy's supra-orbital reflex is present. Deep reflexes are absent.

Sphincters.—While the patient suffered from incontinence of urine up to his tenth year, he can now hold his urine very well, in fact, too well. He has great difficulty in emptying his bladder. Constipation is obstinate.

Psychic Functions.—The intellect is not impaired. He is a good arithmetician, reasons well, and attends to his business without mistakes. His usual cheerful disposition has remained unchanged. Although of late he finds fault with his memory, no sign of psychic deterioration is discernible. About four months ago a peculiar speech disorder developed, which has the attributes of both the paretic and scanning speech, and which is very difficult to describe. When asked

to repeat the usual paradigmata, "Peter Piper," etc., "Round a rugged rock," etc., he shows some degree of syllable slurring. But in conversation he seems rather to scan his words and leaves out no syllables. In the absence of any, even the slightest, psychic alterations, the writer is led to attribute this speech disturbance to the lingual and facial ataxia mentioned above.

April 8, 1904: Patient was suddenly taken ill with symptoms of acute cold. Two days later he developed a right lobar pneumonia and was transferred to Wesley Hospital. During his entire illness the temperature did not rise above 102.4° F. (39.1° C.), but he was delirious most of the time and had to be kept in restraint. There was an uneventful recovery, and the patient left the hospital on April 24th. Subsequently, when visited at his house one week later, patient was in excellent spirits. He said his nerves were steady and he felt strong and well. His demeanor was rather quiet, as were also his facial muscles. The twitching previously observed was reduced to a minimum and even the left-sided ptosis previously noted was ill defined. The tongue was less tremulous than it had been before his acute sickness, and his speech was almost normal. An improvement in the nervous symptoms of patients affected with chronic nervous disease has often been observed after the subsidence of a complicating acute infectious disease.

Simple enumeration of the symptoms leaves no doubt of the diagnosis of *juvenile tabes*, to wit: loss of deep reflexes, ataxia, rigid and unequal pupils, primary progressive optic atrophy, ptosis, lightning pains, bladder disturbances, girdle sensation, paresthesia. One must bear in mind, however, the two disorders for which juvenile tabes has so often been mistaken: *Friedreich's ataxia* and *cerebrospinal lues*.

Friedreich's ataxia, according to Charcot's beautiful lecture, may be clinically thought of as a combination of tabes and multiple sclerosis. In this disease there is the hereditary and family element, the ataxia is cerebellar in type and occurs both at rest and in motion; there is a chorea-like oscillation of head and trunk; the upper extremities are early involved; knee jerks are lost late; bladder disturbance is rare. There are no lancinating pains; sensory disturbances, if present at all, are slight; there is scanning speech, nystagmus, scoliosis, club-foot, paresis in the peroneal region, vertigo and some disturbance of intelligence; no amaurosis, optic atrophy, or Argyll Robertson pupil. It will be observed that neither this description, nor that of others, shows any similarities with this case.

As for cerebrospinal lues, it is certainly possible for that disease to assume the mask of tabes until discovered on the postmortem table; or syphilis and tabes may both be present. Anatomical findings of syphilis of the central nervous system in patients who, during life, presented a typical picture of taboparesis, have been described. There is even difficulty in the correct anatomical interpretation of sclerosis of the posterior columns where luetic spinal meningitis is also present.

To sum up, we have an uncomplicated case of juvenile tabes in

which syphilis is proven in all the relatives. The patient himself has the mark of syphilis on his person, however mild the disease may have been. The postmortem examination of this case showed the typical lesions of tabes.

Treatment.—**PROPHYLAXIS.**—Our greatest hope in the prevention of parenchymatous neurosyphilis lies in an adequate handling of the infection in its early inception. To this end those who see syphilis in its early manifestations, namely, the genito-urinary surgeon and dermatologist, should possess a fair knowledge of the course of neurosyphilis. Syphilologists are now in favor of lumbar puncture being made on patients when they first present themselves for examination at certain intervals during the course, and certainly at the conclusion of treatment. A patient may have become free from subjective discomfort and consider himself cured, when in reality he has not received adequate treatment, for the spinal fluid may still reveal lymphocytosis, an increase in globulin content and a strongly positive Wassermann reaction. Again, the examination of the spinal fluid may reveal a latent syphilitic infection, which if unrecognized, may be permitted to remain in the body for years until awakened to its existence by the apparently sudden development of symptoms of tabes or paresis. Of course, when clinical symptoms have become sufficiently marked for a diagnosis to be made, then the damage to nerve-structures may have become irreparable. Not only has the physician a duty to perform in the matter of prophylaxis of parenchymatous neurosyphilis, but much depends also on *individual prophylaxis*. There are those who have never learned to live according to the rules of hygiene, whose entire life consists of a series of debauches. No wonder that such persons develop tabes or general paresis, once they have become infected with syphilis. If Edinger's theory is correct—that those portions of the nervous system which are used excessively are the ones to be affected—then the logical inference would be to spare the parts subjected to the greatest strain. This should be attempted by the avoidance of excessive walking, running, or forced marches; also by the avoidance of excessive sexual indulgence. In addition, poisons having a tendency to undermine the nervous system, as alcohol and tobacco, must be strenuously avoided. The individual who has once acquired syphilis must consider himself a patient during the rest of his natural life. The physician is not only obliged to treat his early and late symptoms, but must also be on the alert for the first signs of parenchymatous syphilis of the nervous system, of which tabes is the principal representative. Not rarely patients have received treatment only until the blood Wassermann was made negative, without regard to the existence of symptoms in the nervous system, as revealed by clinical signs and corroborated by a positive spinal fluid Wassermann, ordered perhaps by the next physician. The best prophylaxis of tabes seems to be the thorough and prolonged treatment of syphilis, frequently controlled by blood and spinal fluid tests.

What is the principal object in the treatment of tabes? Generally speaking, the retardation or arrest of the disease as a whole. Knowing that tabes in some instances has a natural tendency to become arrested, either temporarily or permanently, we shall be rather conservative in the correct evaluation of any new method of treatment.

Until recently, however, the physician's attitude was only that of a critic and skeptic. It is not so long ago that the physician thought his entire duty was done when he told his patient, after a painstaking examination, that the disease is called locomotor ataxia and is incurable. Until a few years ago tabetic patients were kept in our general hospitals long enough to be diagnosed and presented to medical students for clinical demonstration; then they were either sent home or to the infirmary. All this has changed; to-day we not only diagnose the disease, but we also treat the patient.

The treatment may be conveniently discussed under the headings of (1) causal treatment, (2) symptomatic treatment, (3) mechanical treatment and reëducation treatment, (4) general plan of treatment.

(1) CAUSAL THERAPY.—The real causative factor of tabes is syphilis, but until recently neurologists were not agreed as to the advisability of giving antisyphilitic treatment in cases of tabes. However, even before the Wassermann era and prior to the finding of the spirochetes in the cord by Noguchi there were men like Erb and others with wide experience in neurosyphilis who advised the usual antispecific treatment. At the present time, when there is no more talk of metasyphilis or parasyphilis, and when almost every sophomore speaks of parenchymatous syphilis of the nervous system, there is no excuse for withholding antisyphilitic treatment from a case of tabes. Of course, there are contra-indications for the administration of the causal treatment; cachectic patients and those suffering from advanced arteriosclerotic and nephritic changes are not fit subjects for the intensive treatment by means of mercury and salvarsan. In the writer's opinion, every case of tabes should be given an opportunity to receive the possible benefits from the modern treatment of parenchymatous syphilis.

The drugs utilized in the treatment of tabes are: (a) **iodids**, (b) **mercury** in its various forms, and (c) **salvarsan** and its equivalents.

(a) *Iodids*.—Few modern authors continue to advise the use of iodids in the treatment of tabes. The writer has learned to regard the iodids as a drug which deranges the patient's digestion, thus reducing his vitality without adequate compensation in the way of benefit for his disease. Its use, therefore, in therapy of tabes has been practically discarded by the writer, who retains it only for that group of cases belonging to the exudative type of the disease, in which doses of from 15 grains (1.0 gram) to 1 dram (4.0 grams) are given three times daily after meals.

(b) *Mercury*.—The administration of mercury not only in so-called tonic doses, as advised by the older writers, but in massive doses, the same as for interstitial syphilis, has become a recognized form of treating tabes. It is given either alone or in conjunction with salvarsan.

Directions for its proper administration will be found under Treatment of neurosyphilis (pages 124-125).

(c) *Salvarsan*.—Patients are given salvarsan intravenously, either alone or in combination with mercurial ointment, rubbed into the body, or mercurial injections are administered intramuscularly. The salvarsan or one of its equivalents is injected intravenously once a week, while the mercury rubs are used nightly. A series of a dozen treatments may be followed by a three weeks' rest; then another such series may be introduced. After six treatments the spinal fluid and blood should be examined for Wassermann. The intravenous salvarsan and mercury treatment may be of great value and perfectly adequate. Symptoms are relieved and the patient may show general improvement. There are many patients, however, who have been given dozens of injections with only indifferent or no results. Such cases should be given the benefit of intraspinal injections according to Swift-Ellis. The writer has seen beneficial results from this method beyond his most sanguine expectations, which were denied him by the simpler therapy. Patients who suffered from excruciating pains and most atrocious attacks of gastric crises were relieved by intraspinal injections, after fifty or more intravenous injections failed to produce any appreciable effect either on the clinical symptoms or on the serological picture. Of course, the writer is far from deluding himself into the belief that he has found a curative remedy in autoserosalvarsan spinal injections, but believes it to be the best causal therapy for many forms of tabes. The utmost we can possibly hope for is relief from the distressing symptoms and an arrest of the progress of the disease. The technic of the Swift-Ellis treatment has already been described, and the reader is referred to the section on Treatment of neurosyphilis (p. 120). Here it is sufficient to emphasize its therapeutic value and to enumerate the several methods of treatment.

(2) SYMPTOMATIC TREATMENT.—Perhaps the symptoms demanding relief most urgently are the *lancinating pains*. They may be severe and resistive to any form of therapy except **morphin**; but this should be reserved as a last resort. In a disease in which pains are apt to recur so frequently the administration of morphin means the certain creation of the morphin habit, which, in many respects, is worse than the disease itself. We should, therefore, endeavor to give relief by means of the various coal-tar derivatives, such as **phenacetin**, **antipyrin**, **acetanilid**, **pyramidon**, **aspirin**, either singly or in combination. The hypnotics, **sulphonal**, **veronal** or **luminal**, may be administered with analgesics. For the pains **prolonged warm baths** (98°-102° F.) (36.7°-39° C.) have been recommended, also the application of **dry cups** to the spine and the actual cautery. In a few instances the author has seen relief from pains and gastric crises by the administration either hypodermically or by mouth of 10 minims (0.6 c.c.) of the 1:1000 solution of **adrenalin**, repeated every 2-3 hours. Sicard and Lemoyez have treated tabetic pains by means of intravenous injections of adrenalin, giving one-half mgm. of adrenalin in 10 c.c. of serum. They report

complete cessation of pains within one-half to two hours. The use of morphin is permissible only when the patient resists all other treatment and exhaustion from excessive pain is threatened. It must always be remembered that he who leaves a hypodermic syringe with a patient has thereby made him a morphin habitué. If any morphin is to be administered, it must be given by the physician and only when absolutely required.

Gastric crisis is a symptom which calls loudly for symptomatic relief. Patients vomit for days and come near the verge of collapse unless relieved. The problem is how to check the attack without having redress to morphin. One may try **oxalate of cerium** in doses of 5–10 grains (0.324–0.65 gram), or 10 minims (0.6 c.c.) of **adrenalin** (1: 1000) may be given a trial. In quite a few cases the author has seen relief ensue after two or three doses of adrenalin one hour apart. No rational explanation is attempted for this phenomenon, but the more extensive use of adrenalin in gastric crisis is urged. During the continuance of a gastric crisis the stomach will not retain food, and it becomes necessary to resort to rectal feeding.

For the *vesical crises* **morphin** may have to be injected, especially in the beginning. After this, Collins prescribes a mixture of **chloral hydrate**, **fluidextract of belladonna**, and **fluidextract of hydrastis** in the usual doses.

Laryngeal crises are usually relieved by the inhalation of **chloroform**, which must not be carried to the point of complete narcosis. In the majority of cases morphin becomes necessary, and absolute quiet is a measure automatically adopted by the patient.

Retention and incontinence of urine must be treated by regular catheterization and washing out of the bladder with weak **boric acid** or **permanganate of potash** solutions. **Urotropin** in 15-grain (1.0 gram) doses is given three times daily and also a mixture containing **belladonna** with **fluidextract of hydrastis** or **ergot**. R. Stintzing recommends **galvanization of the bladder** through the abdominal walls in the following way: One large electrode, 6–12 cm., should be placed above the symphysis, and the other electrode of half the size and with a concavity should be so placed that it fits up close beneath the public arch; then a current of from ten to twenty milliamperes should be allowed to flow through from three to five minutes.

Blindness caused by optic atrophy is unfortunately one of the symptoms least amenable to any sort of treatment. However, for patients with only partial blindness it has been the author's custom to resort to the most vigorous **combined salvarsan and mercury treatment**. If after two months of such treatment there is neither improvement nor aggravation of symptoms, the mercury is discontinued and a course of **intravenous-intraspinal** (Swift-Ellis) **injections of salvarsan** or its equivalents is begun. The patient receives one dozen injections at intervals of ten days between each. In addition, the methodical ad-

administration of **strychnin nitrate** hypodermatically is begun, in doses of grain $1/30$ (0.002 gram), increased to grain $1/20$ (0.003 gram), twice daily. This treatment may be continued for months. While continuing the administration of strychnia, the Swift-Ellis courses of treatment, consisting of one dozen injections, are repeated, with intervals of three months between each course. Those who are treated at home and who have no nurse at their disposal are given **strychnin sulphate** grain $1/20$ (0.003 gram) by mouth, which dose is gradually increased to grain $1/12$ (0.005 gram) of **strychnin sulphate** three times daily. It is surprising what large doses of strychnia can be tolerated by patients, provided the doses are increased gradually. From the treatment outlined the writer has seen arrest of the blindness which he is loath to attribute to spontaneity, though it must be admitted that there is no positive proof to that effect. On the other hand, some cases go on toward inevitable blindness, regardless of what treatment is pursued. Optic atrophy blindness is perhaps the saddest lot of the tabetic. It cannot be denied, however, that while this is hard to bear, yet the tabetic who develops optic atrophy early in his disease seems thereby to remain immune from the many disagreeable spinal symptoms of tabes. There is no satisfactory explanation for this phenomenon, but the fact is indisputable.

Bed-sores are much more easily prevented than cured. Every possible precaution should be taken toward that end. The parts which are subjected to continual pressure should be well padded and the skin and subcutaneous tissues must be kept clean by **daily cleansing baths** and frequent **sponging with cold water and alcohol**. Immediately after bowel movements and after each urination the parts should receive careful attention, as bed-sores are mostly found on portions of the skin which have been allowed to come in contact with the excretions. When bed-sores have already formed, they must be treated by **strictly surgical means**. The same applies to *perforating ulcers of the foot*.

Regarding the *arthropathies and osteopathies*, **orthopedic measures** will find their useful application in these and the other trophic disorders of tabes.

(3) **MECHANICAL OR PHYSICAL THERAPY**.—The measures included under this heading are **hydrotherapy, electrotherapy, operative procedures**, and **Frenkel's reëducation methods for ataxia**.

(a) **Hydrotherapy**.—Tabetics occasionally derive much benefit from hydiatic measures when applied to suit the individual case. The effects are both tonic and symptomatic. Individuals who have lost weight and who suffer from insomnia due to pain may receive great benefit from a course of systematic hydrotherapy. This may be obtained in many European and American watering places, whence tabetics return in better health, partly because of the complete freedom from worry and work and partly because of the beneficial effects of the treatment. Local hydiatic measures are applied for the relief of tabetic pains and also for the urinary disturbances. As patients are usually referred for such therapy to hydrotherapeutic establishments, it is unnecessary

to go over details which are generally arranged by the hydrotherapeutist in charge of the institution.

(b) *Electrotherapy*.—The value of electricity in the treatment of tabes is differently estimated by various authorities. It is true that formerly electricity was accorded an important place in the treatment of tabes. Some authors in Europe and America have lauded it to the skies, while others denied it any therapeutic virtue. There are those who believe that the benefits, if there are any, are caused purely by suggestion, and cite as proof the statement, that any new or unusual method of treatment appeals favorably to a tabetic. Whatever may be the explanation, the fact remains that some tabetics appear to benefit by the galvanic current applied to the spine and given either with the labile or stabile electrode. The writer believes too much space is usually devoted to a description of the exact application of the current to the spine; he has found it beneficial to apply as much current as the patient can comfortably stand—one sponge electrode over the cervical, the other over the sacral end of the spine. As the efficacy of this treatment may be entirely due to the psychical effects, the patient should at least be made to feel some pain where the current is applied.

The paresthesia of tabes has in many instances been relieved by the application of the faradic brush.

Massage is used in conjunction with electricity so as to maintain muscular tone and thus improve the general health of tabetics who are unable to move about.

Though neurologists generally have somewhat neglected physical therapy and especially electrotherapy in tabes, general practitioners have been too sanguine in their expectations and have thus led patients to neglect more rational means of treatment. There is no doubt, however, that the judicious application of electricity is helpful in tabes.

(c) *Operative Relief of Gastric Crises*.—Section of the posterior roots—posterior rhizotomy—was first suggested by Förster, in 1908, on the theory that the severe gastric pains in the attacks were caused by irritation of the cell-bodies of the sensory sympathetic fibers of the stomach. These pass from the celiac plexus in the great splanchnic nerve to the cord through certain of the posterior roots. If this view is granted, the crises should cease after the sensory pathway from the stomach has been intercepted by the severance of the posterior roots involved. The first operation of this kind was performed, in 1908, by Küttner upon one of Förster's patients. The sixth to the tenth posterior thoracic roots were sectioned and the pain and vomiting ceased. Since then the operation has been done repeatedly with varying degrees of success. About half of the operated cases have been either partially or entirely relieved of the unbearable pain and incessant vomiting. Frazier, in "Surgery of the Spine and Spinal Cord," after discussing the various surgical methods introduced for the relief of gastric crises, gives an excellent summary, a part of which is herewith quoted as representative of the latest thought on the subject. He says:

"With or without a positive Wassermann reaction every case without exception should be given vigorous antisyphilitic treatment before the question of operation should be dreamed of. In the event of failure to relieve suffering, what then? Alcoholic injections are in the developmental stage and cannot now be endorsed. Beginning with peripheral procedures, Jaboulay's operation, stretching of the solar plexus, a fanciful idea, does not deserve consideration because of inevitable recurrence. Franke's extraction of the intercostal nerves, through an extraspinal operation, has been attended with a high mortality. It is a hazardous and crude undertaking which may carry away both anterior and posterior roots and tear the dura. Recurrence is the rule and the longest period of relief eleven months. Approaching the cord, there comes for consideration the extradural gangliectomy of Sicard and Desmarest, an operation applicable only to lesions of the thoracic region without sufficient clinical application to warrant endorsement. Theoretically, ligation of the anterior and posterior roots, proposed by Sauv   and Tinel in 1913, appears to me as meritorious. The operation has not been tried and essentially differs in no respect from division of the posterior roots. Either intradural or extradural rhizotomy, preferably the former, is in the judgment of the writer the operation of choice. If undertaken at all, one should not wait until the patient is physically unfit as an operative subject, and at least as many roots as are represented in the area of hyperesthesia should be divided, usually five or six. If there be an extension of the crises beyond the original zone, two or three more roots may be divided at a second operation. A fair appraisal of the results of intradural rhizotomy may be expressed in these terms: *two in ten cases cured, four in ten improved, one in ten unimproved, and one in ten die.*"

(d) *Re  ducation of Ataxic Patients—Frenkel's Method.*—This method aims to treat one symptom only, namely ataxia, and does not claim to exert an influence over the entire course of the disease. Fortunately not all tabetics become ataxic, but those who become dependent upon crutches or the support of attendants, and are, toward the last, bed-ridden, deserve our commiseration. The re  ducation method is a great boon to such patients, for by such means many of them have again become able to enjoy the pleasures of life after having been invalids confined to their beds.

In 1889, Frenkel elaborated his system designed to re  establish coordinated movements in tabes patients. He began with an extensive study of the true nature of ataxia and hypotonia which was followed by therapeutic hints. The essentials of his treatment, scattered in various publications, subsequently appeared in one volume, translated into English, in 1902, under "Tabetic Ataxia." The book is worthy of perusal by all those who are concerned with the treatment of tabetic ataxia, and the following directions are largely abstracted from it. Frenkel's exercises have the merit of being simple and adaptable to the needs of various degrees of ataxia. Though he devised a number

of apparatus, many of the exercises can be carried out without them. Essentially these exercises are directed to the redevelopment of correct coördination in the upper and lower extremities. It is a system of "cerebral gymnastics," rather than one of "gymnastics by force," as Frenkel characterizes Leyden and Goldscheider's somewhat similar exercises.

Frenkel's procedure consists in teaching the tabetic to perform exact and methodical movements requiring skill, not force. In the beginning the movements are simple and elementary and, as the patient acquires efficiency, they become more complicated. The exercises are best done under the direct supervision of a physician or a specially trained attendant, for method and system are essential to success.

As in other methods of treatment, there are indications and contra-indications. Generally speaking, this treatment is indicated whenever ataxia appears early and develops rather slowly, provided the patient has not entirely lost his deep sensibility. The contra-indications apply to those cases in which the individual has become weak and cachectic, or suffers from frequent attacks of gastric crises. Those suffering from the acute and subacute forms of tabes are not fit subjects for Frenkel's exercises; neither are the patients suffering from trophic changes of bones and joints, and thus liable to spontaneous fractures. Likewise, those affected with optic atrophy, which latter condition appears to retard the approach of ataxia, are usually excluded from the exercises. The work is best carried out under supervision and in groups of three to five patients.

The patient is encouraged to practice those movements, motions and steps in which he is particularly deficient. By persevering in these exercises many a bedridden patient has again learned to walk.

The exercises are of two classes, those performed in bed and those performed out of bed, depending upon the patient's disability. The following is quoted from Church and Petersen, who have made a concise résumé in English of Frenkel's exercises for tabes:

"In bed, the patient is called upon to flex, extend, abduct, and adduct each leg separately and then both simultaneously. The knees and hips are likewise exercised. The patient is asked to place the heel of one foot on the big toe of the other foot. Place heel upon knee of the other leg and then slowly travel along the ridge of the tibia toward the ankle. Exercises are made alternately, first with one leg, then with the other, with open and with closed eyes. These exercises are attempted over and over again, with frequent rests. Patient is encouraged to persevere until he succeeds.

"The exercises are repeated twice a day, a half hour in the morning and again a half hour in the afternoon.

"1. Patient is placed with his back to a chair, heels together, then seats himself slowly in the chair, and is then made to rise in the same careful manner. No cane is used. If patient cannot stand, an attendant is placed on either side to support him if necessary.

"2. One leg is placed at an ordinary walking step in front of the

other, and then placed with great exactness back into its original position. Same exercise is then performed with other leg. The patient, if necessary, supports himself by a cane or otherwise.

"3. Walks three paces slowly and with precision.

"4. Rest in standing position, one foot before the other; with arms placed akimbo, he flexes his knees and slowly raises himself again.

"5. Patient, as in exercise number 2, advances one foot, then returns it to its original position, and then places it one step behind the other. This exercise is usually a very difficult one, requiring, as it does, a great deal of balancing power.

"6. Walk twenty steps, as in exercise number 3.

"7. Number 2 performed without a cane.

"8. Stand without a cane, with feet placed together and hands on hips.

"9. Stand without a cane, feet separated; various movements with the arms, grasping objects, forcing back outstretched hand of physician, etc.

"10. Maintain same position as in number 9, flexing trunk forward, backward, to the right, and to the left.

"11. Exercise number 9 with the feet together.

"12. Exercise number 10 with feet together.

"13. Walk along a painted line on the floor, patient supported by a cane.

"14. Same without a cane.

"Exercises for the fingers and arms are also employed, based on the above-mentioned principles.

"These various exercises are to be progressively attempted and persevered in as coördinate strength improves. They may then be gone over again with closed eyes aided by a cane or assistant, then without aid. Fatigue, however, must be avoided. In advanced cases that have to rely on crutches, a tall 'walking-frame' or roller-crutch, such as is used for children, may be employed. This gives support under the arms and enables the patient to exercise the legs. Precise, delicate motions with the fingers may also be developed in the same way."

(4) PLAN OF TREATMENT.—Shall we inform a tabetic of the true nature of his malady? This is a question to be decided immediately after the diagnosis has been made. In the writer's opinion it is best to explain to the patient that he suffers from an organic disease of the spinal cord which requires energetic and continuous treatment. It is immaterial to the purpose whether one names the disease or explains its causation—the only thing needful is to enlist the patient's coöperation. This information may startle the patient, but after the first few minutes he becomes reconciled to his fate and begins to ask questions about definite arrangements for treatment. Without a knowledge of the true condition, it can hardly be expected of any one to subject himself to the expensive, tedious and often painful treatment required for the average case of tabes. In an advanced case of tabes the patient

usually brings the diagnosis with him and is chiefly interested in the possibility of obtaining amelioration of his symptoms.

In the treatment of this organic disease **psychotherapy** is by no means to be neglected. It is this neglect of the mental element in the treatment of all forms of nervous disease, functional and organic, that is largely responsible for the rapid growth of the various "pathies." If the quack knows nothing of scientific medicine and diagnosis, he is sufficiently acquainted with the psychic end of it to outdo the average practitioner. We must recognize facts as they are and not as they should be. What are the facts? Tabetics need constant encouragement and optimistic suggestions; their spirits must be raised from the depressed moods into which they habitually sink. Maloney is correct when he makes the statement: "With equally extensive lesions the brave ataxic walks, the timorous is bedridden; a slight urinary disorder or a loss of sexual power may in one tabetic scarcely scratch the mental surface and in another produce intolerable disease; one bears cheerfully slight dullness of hearing, which in another becomes an isolated deafness. The mental influence varies also with the nature of the symptoms." But this author is wrong when he speaks lightly of the entire drug-treatment of tabes, by saying: "The circumstances attending salvarsan injections, together with that drug's reputation, is responsible for much of the alleviation of pain, fatigue and depression which sometimes follows its use; and this psychic effect is especially marked after the rubric which attends the intraspinal administration of salvarsanized serum." And, further, while it may be true that "neither the transient inhibition of the growth of the spirochetes nor the indiscriminate general excitation of the vegetative nervous system, exerted by the metallic substances which we now employ as antisypilitic remedies, but the development and reënforcement of the natural human defense against the spirochete by well-directed stimulation of the vegetative nervous system, with specific, unorganized, chemical substances, such as vaccines, will ultimately afford the cure for which we seek," there is no likelihood that this will soon be realized. Indeed, it is doubtful if the line of therapy now followed will ever be changed to a radically different one. There surely will be improvement and change of method, but the writer feels confident that we have not adopted another one of the numerous psychotherapeutic measures when we subjected our patients to the ordeal of intraspinal therapy. For this purpose there are many methods easier and less costly than the intensive mercurial and salvarsan treatment.

Having made his viewpoint clear as to the need of optimism in the handling of tabetics, without allowing himself to be deluded into a belief that everything done for the patient is the result of psychotherapy, the writer will mention a few of the leading principles.

All treatment must be supplemented by hygienic, dietetic, physical and disciplinary measures. Correct treatment no doubt has a tendency to prolong life; the sooner it is instituted the better for the patient. Early recognition means, that when a patient consults his physician for

any ailment whatsoever, tabes should be thought of, especially when there is a history of syphilis.

The neglect of this rule is demonstrated daily when we see patients who have wasted time and money with genito-urinary surgeons, ophthalmologists, and general surgeons, for the treatment of diseases supposed to belong to their respective specialties. Many are the ones who have been operated for gall-stones, appendicitis and intussusception of the bowels when they were really suffering from gastric or intestinal crises. In any case presenting vague symptoms and a doubtful diagnosis it is our duty to insist upon a spinal fluid examination, the result of which may furnish not only diagnostic aid, but may also serve as an index to treatment.

The question frequently asked by patients as to the necessity of a change of occupation leads to the following considerations: Generally speaking, patients should not be permitted to become semi-invalids by being ordered to give up their occupation. Such advice is harmful, because idleness in tabes is conducive to introspection, hypochondriasis and constant brooding, all of which cause the patient's energies to fritter away, while those patients who are regularly occupied seem to fare better. Tabetic patients should be permitted to spend a portion of their time in business or professional work, the rest of the time being devoted to what may be called recreational treatment. This will consist at first in **walking, driving, golfing**; followed by the judicious application of **hydrotherapy, electricity, massage and rest**. In the writer's opinion, sanatorium treatment is not to be recommended, except for those tabetics who cannot receive proper care at home and for those who have reached the last stages of the disease. As patients are easily affected by even slight changes in temperature, a sojourn in a mild climate during the winter is beneficial.

Regarding the **diet, tea, coffee and stimulants** are **absolutely prohibited**; a **mixed diet** of vegetables, fats, and proteids in proper proportion is the ideal one. The patient should rest in bed twelve out of the twenty-four hours. Ordinary exercise is good for the tabetic, but he must be warned against excessive fatigue. Sexual indulgence is either to be entirely prohibited or must be reduced to a minimum. No disease requires greater **attention to the details of general hygiene**, such as care of stomach and bowels and of the skin by baths. Those suffering from lack of appetite and deficiency of hydrochloric acid should be given **stomachics** with **hydrochloric acid**.

For the *nervousness and restlessness in tabes* the author prescribes **sodium bromid** in moderate doses, 15 grains (1.0 gram) three times daily after meals. **Strychnia**, which has long enjoyed the reputation of being a good nerve-tonic, **should not be prescribed** in tabes, for it may cause hyperirritability and sleeplessness, as well as an increase of lightning pains and irritation in the urogenital sphere.

Prognosis.—The outlook as to life is good, provided the diagnosis has been made early and proper measures have been instituted to combat the disease causally and to relieve symptoms as they occur. Of

course, there are types of the disease which may properly be called fulminant and whose tendency is to materially shorten life. Fortunately these are the exceptions. On the whole, the prognosis as to life may be considered favorable, though it must be stated that tabetics have a lessened power of resistance and easily succumb to infections, particularly tuberculosis. Quite a few are carried off by chronic septic poisoning from bed-sores, cystitis, pyelonephritis, and general cachexia.

Regarding optic atrophy and blindness, it may be stated dogmatically that no amount of treatment is capable of restoring vision, but our efforts are often successful in arresting further progress of the condition. One is never certain, however, whether the treatment is solely responsible for such arrests, or whether these are spontaneous remissions, as even without treatment optic atrophy may remain non-progressive during a period of years. Much has been written on the tendency of mercurial and arsenical preparations to cause an aggravation of eye symptoms and to produce optic atrophy, but the writer believes we are now learning that there is no real foundation for such reasoning. The explanation may be that the atrophy had been progressing so rapidly that not even treatment could stop it. Much has also been written on the subject of neuro-recurrences, which means that salvarsan or mercury treatment supposedly produce lesions in cranial nerves not originally present in these cases. Most of these discussions are now obsolete, for it has been repeatedly demonstrated that cases in which there occurred cranial nerve accidents were either treated with toxic salvarsan or the injections were improperly made. The writer has never seen any accidents from the treatment of tabetic optic atrophy. On the other hand, several patients have been treated by him who were decidedly benefited by the mixed salvarsan and mercury treatment; in two instances results did not appear until the Swift-Ellis treatment was resorted to. The prognosis in optic atrophy, while inherently bad, is by no means made worse by treatment, but may be considerably benefited thereby. Certain it is that though in the majority of cases optic atrophy seems to protect the patient against the advent of the more serious symptoms of tabes, we must not remain indifferent to the progress of the entire disease by assuming an attitude of inactivity. It is obvious that treatment is absolutely necessary to influence the prognosis.

Regarding the return of the tendon and of pupillary reflexes, though some have reported this occurrence, a return of reflexes, once they were completely lost, has never been seen by the writer. In cases with sluggish response, he has seen what he thought was improvement.

The prognosis as to subjective symptoms is more favorable. After treatment the lancinating pains and disagreeable paresthesiæ have shown decided improvement. The marked incoördination is also favorably influenced by causal treatment independent of the Frenkel exercises. In the case of one patient, completely ataxic and unable to leave his bed, it has been the writer's privilege to see him walk after five Swift-Ellis treatments. While specific treatment exerts a favorable influ-

ence on incoördination, the greatest triumphs in ataxia are obtained through the Frenkel treatment. Patients who navigated about with the aid of two canes were able to walk without support; and even bedridden cases have been converted into ambulating individuals after persistent efforts with the Frenkel method.

Pathology.—The pathological changes are found principally in the posterior roots and the posterior columns, though the sensory spinal nerves also show degeneration, greatest at the periphery, and mostly limited to the sensory fibers for skin, muscles, and joints. Upon opening the dura of the spinal cord, the membranes are found thickened over the posterior surface and the cord itself appears flattened and gray or grayish-red. The posterior roots, normally two or three times as large as the anterior ones, are also thinned out, wasted, and considerably smaller than the anterior roots. Upon sectioning the cord the posterior columns are found to be shrunken and grayish as compared with the anterior and lateral tracts. The greatest amount of degeneration is observed in the lumbosacral region, diminishing from below upward. The degeneration does not appear limited to the sensory neurons of the spinal cord, but may also implicate the special sense neurons, as those of vision and of hearing.

The *microscopical examination* of the cord reveals nerve fibers in which the myelin sheaths are diminished or destroyed, and the axis cylinders appear irregular in thickness or have been completely destroyed. Everywhere neuroglia has increased, while the parenchyma itself has become lessened. Generally speaking, the exogenous fibers—or those entering the cord from without—are destroyed, while the endogenous fibers—or those arising from cells within the cord—may remain intact. The arteries and veins situated in affected tracts are usually found sclerosed. In addition, the pia-arachnoid is in a state of chronic inflammation and considerably thickened.

And in like manner the intervertebral or posterior root ganglia are found reduced in size and number, and nuclear displacement and granular changes are often encountered in their cells. The ganglion cells being trophic centers for the sensory nerves and the posterior root fibers, their pathological changes may explain many of the sensory phenomena of tabes. Degeneration of the visceral sympathetic nerves has been described, the degenerated fibers having been followed into the cord.

The cord lesions may perhaps be explained by a review of the anatomical distribution of the posterior roots. It is to be recalled that the posterior roots divide into ascending and descending fibers. The ascending fibers again divide into (*a*) a tract of fine fibers running directly upwards to the tip of the posterior horn, which receives many of the fibers. This is Lissauer's tract, which may degenerate early in tabes. The fibers of the second group (*b*) pass to the inner side of the first, forming what is known as Burdach's columns; these enter the middle third of the inner border of the posterior horn. Of the fibers some enter the gray matter and arborize around cells found

therein; others come into relation with the large cells of the anterior horns, with cells of the posterior horns, and possibly with those of the intermediolateral tracts.

The loss of the deep reflexes and of muscle tonus is probably the result of degeneration in the posterior root fibers which terminate in the anterior horns, the reflex arc being broken at this point. Another set of fibers of medium length (*c*) ascend for a certain distance and then enter the gray matter, to be distributed as terminal fibers around the cells of Clarke's columns, which latter are known to give rise to the ascending cerebellar tracts. Degeneration of this tract, common in tabes, may explain the disturbance of equilibrium. A fourth group of fibers (*d*) enters near the median line and passes up almost the entire length of the cord, forming what is called Goll's columns, which eventually terminate in the nucleus of gracilis and the nucleus cuneatus, whence originates a new sensory path to the cerebrum. The long fibers, as they ascend, are gradually pushed over toward the median line and backwards by the nerve-roots above them which enter in the same way, namely, at the inner border of the posterior horn. Thus it happens that the fibers from the lower part of the cord occupy the posterior median columns and those which come in at higher levels occupy more lateral positions and practically constitute the column of Burdach. From what precedes it will become evident that the pathological appearance of tabetic cords will differ in different cases according (1) to the position of the nerve-roots involved, (2) number of roots implicated, and (3) completeness of degeneration. Thus in tabes affecting the upper part of the cord, cervical tabes, we find in the cervical region the columns of Goll healthy, but Burdach's columns show degeneration, while in the lower part of the cord the posterior columns are healthy in their whole extent. In the usual type of tabes, the dorsal and lumbar regions of the cord show extensive degeneration of the posterior columns, involving the entire cross-section, while in the upper parts of the cord with the incoming healthy nerve-roots, only the columns of Goll will be degenerated. And lastly, it may be mentioned that there is never complete degeneration of all the sensory tracts in the cord. Even in the most advanced case of tabes certain small tracts of fibers may remain intact, namely those fibers which are considered to be "endogenous," that is, which originate within the cord itself and are not prolongations of the nerve-roots entering from without.

Where does the degeneration begin? It is natural that it should begin in the ganglia of the posterior roots, since the cells of these ganglia are the trophic centers of the whole posterior root system. Yet these ganglia are not always affected—some observers have found slight changes, others none.

Some authors believe the spinal degeneration begins in disease of the meninges, affecting the spinal roots at certain points where they are peculiarly vulnerable. According to Nageotte, one such point lies where the posterior root pierces the dura mater and receives, in common with the closely adjacent anterior roots, a sheath of connective

tissue from the dura and arachnoid. A chronic meningitis might give rise to a transverse neuritis at this point, and so start a degeneration in all the ramifications of the nerve-roots. The objections to this hypothesis are, first, that the existence of such a meningitis—at least as a necessary antecedent of tabes—has never been proven, and secondly, that the adjacent anterior roots would almost certainly be involved as well, so that muscular atrophy in tabes ought to be the rule instead of the exception. Redlich and Obersteiner's theory puts the vulnerable point at the entrance of the posterior root into the spinal cord. Here even normally the pia mater forms a constriction around the entering nerve-root and causes a narrowing of its caliber, so that if any inflammation of the pia took place, it might result in additional constriction, thus injuring the nerve-root and consequently its intraspinal ramifications. Of course, the hypothesis shares the same weakness with Nagotte's theory in that the meningitis as an early occurrence in tabes has not been proven.

It is well to note, however, that at this point the nerve fibers lay aside their neurilemma sheath. And whether it is because of the denudation of neurilemma or, as Marie and Guillain maintain, because of the peculiarities of the lymph circulation of the posterior columns which bring it here into special relation with the lymphatic spaces of the meninges, it seems probable that the point of entry of the posterior roots may afford a ready channel for the spirochetes entering the cord. What we actually notice in tabes is the gradual death of the neuron, beginning in the branches of the tree, usually the intraspinal branches, but sometimes also the peripheral branches, and gradually reaching the cell, which is their root. But other parts of the nervous system are affected which cannot be explained by any of the above theories, as for instance the optic and auditory nerves as well as the trigeminus. And again there are slight changes in the cerebral cortex, corresponding to slight mental involvement, and there are the striking instances in which long-standing tabes terminates in general paresis. It would appear that the morbid process in tabes is of a general nature and of wide distribution rather than a local and accidental lesion of the posterior nerve roots. These are the older views on the pathogenesis of this disease. It is now necessary to state what appears the probable pathology of tabes in the light of the discovery of the spirochetes as the actual cause of all forms of syphilis.

It seems plausible that the starting-point of tabes is an exudative change which is now considered of an inflammatory, not of a toxic origin. Noguchi and others have found spirochetes in the tabetic exudate. The real cause of tabes is no doubt the *Spirochæta pallida*. As a positive Wassermann may be detected in the blood only a few weeks after the primary infection, and as, according to numerous observers, a majority of the cases of syphilis in the so-called primary and secondary stages of the disease show evidence of infection in the cerebrospinal fluid also, it must be inferred that the local lesion of syphilis is really the beginning of a general infection—a thought already expressed by

the older pathologists. The theory has been advanced that the spirochete assumes certain characteristics, depending upon where it has been domiciled. Thus, after a sojourn in the brain of one individual, the spirochete affects the brain of the next individual when transferred to him by infection. The spirochete grows best in the medium whence it is derived. The strain of the spirochete is somewhat specific for the tissue which it invades. There may be several separate strains of spirochetes, some producing skin lesions, others invading the deeper tissues and producing tabes and central nervous system lesions. This may explain why tabetics and paretics have suffered from such mild forms of skin syphilis. Numerous instances have been reported of syphilis acquired from one source by a number of individuals, which has produced the same kind of lesions in all.

In order that the nervous system shall become affected, there must be present a strain of spirochetes with a preference for the nervous system and, *in addition*, poisons which undermine the resistance of the nerve elements, and perhaps occupational influences which render the nervous system most vulnerable.

In the central nervous system the spirochete may attack indiscriminately either the cord or brain, or any portion of the central nervous system. Thus, optic atrophy is often followed by tabes and is found as part of tabes. Likewise there is a close relationship between tabes and general paresis. The cranial nerves have frequently been found implicated; also the cerebellum. In addition the spirochetes have been found in the lymphatic channels of peripheral nerves. It may be a fact that fatigue of certain parts of the nervous system has much to do with the localization of the spirochetes in these parts. According to Edinger, the neurons exhausted by over-function resist the invasion of the spirochetes less than those not so fatigued. As the sensory neurons of the posterior roots are almost incessantly in operation, they are the first to become implicated in the specific process. Further, it appears that those portions of the nervous system are first attacked which offer least resistance. If an optic nerve is first to be attacked, the damage is usually more severe than if the optic nerve lesion follows the cord lesions. The spirochetes may attack either in rotation or simultaneously various parts of the nervous system, which explains the different modes of the onset of symptoms in tabes.

(b) GENERAL PARESIS

Etiology.—The cause of general paresis is syphilis. "No syphilis, no general paresis" has become an axiom. The findings by Moore and Noguchi of spirochetes in paretic brains have demonstrated the close causal relationship between the microscopic organism and the lesion. Nevertheless there are differences, both pathological and serological, between this disease and the earlier forms of neurosyphilis belonging to the interstitial group, which differences have already been dealt with. The disease appears to affect syphilitic brain workers and intelligent

persons more frequently than it does manual laborers and the ignorant. Indeed, it has been termed a disease of modern civilization, for savages have much syphilis and but little paresis. Wherever syphilis is prevalent, general paresis is also found. Fortunately not every syphilitic develops paresis; the proportion is variously estimated, from 2 to 5 paretics to 1,000 syphilitics. In America the disease comprises from 5 to 8 per cent. of the admissions to insane institutions, while in Europe the percentage is many times that number. Army officers are more likely to develop paresis than privates, prostitutes more so than other women, men more often than women. In the form known as conjugal paresis husband and wife may develop the disease at different times; usually the husband seems to suffer first.

Though the real cause of paresis is syphilis, excesses of all kinds, combined with worry and over-work, as well as the exigencies of warfare, are contributory factors. It will be found that the War has multiplied the number of paretics among those who have been the subjects of latent syphilis. A remarkable fact in connection with the development of paresis is the almost universal history of a very mild attack of syphilis. This may account for the previous lack of treatment admitted by nearly all patients; the infection having been mild, there was no indication or desire to pursue systematic courses of treatment. General paresis usually makes its appearance ten to twenty years after the primary infection, and in the form known as juvenile paresis the disease may become manifest between the ages of ten to twenty years.

Symptomatology.—In this type of parenchymatous syphilis there is found a combination of nervous and mental symptoms. The strictly neurologic signs are sometimes in the ascendency, but most often mental signs constitute the leading feature. Dependent upon their habitual temperament, patients become unstable, depressed, exalted, or hypochondriacal. Those of a serious turn of mind will probably show signs of depression, others of the volatile type become exalted or maniacal, and lastly, those of the complaining kind may manifest hypochondriacal tendencies. Because of differences in the symptomatology, four types of cases are recognized: the *demented*, *expansive*, *agitated*, and *depressive*—a classification which is convenient for purposes of clinical description.

Before describing these types it is necessary to give a general outline of the symptomatology, mental and neurologic, common to all of them.

MENTAL SYMPTOMS.—*Clouding of Consciousness.*—Perhaps one of the earliest signs in those suffering from the disease is an increasing difficulty in apprehending clearly the facts of their environment. This refers to a proper comprehension of the finer details in their business or professional lives as well as their domestic relations. There develops what might be called a clouding of consciousness, that is, a condition or state of mind much like that of the dreamer or of the alcoholic while semi-intoxicated. Soon disorientation increases, and the

patient, seemingly rational, neither knows where he is nor what takes place in his environment. He may not be able to name the day of the week, month, year, or season. Indeed, while looking out of the window and witnessing a heavy snowfall, he may answer the examiner's questions regarding the season of the year, by saying that it is summer.

Forgetfulness.—Another early sign is forgetfulness. This is differently shown in the various avocations. The commercial man in his daily business transactions begins to omit many important details. The superior employee—superintendent or foreman—who has no one to control him, may not be noticed to suffer from loss of memory until the disease has made severe inroads, but one who occupies a subordinate position requiring a certain amount of mental labor is more apt to be recognized as showing mental failure. Mechanics and skilled workmen gradually decline in their efficiency both in the matter of quality and quantity. This is according to a law that the highest and most recent acquisitions are the first to disappear in this disease. Early in the disease the memory for recent events becomes defective, which is demonstrated by the patient resorting to written memoranda for everything he wishes to remember, or by adopting other devices for memory-strengthening. As the disease progresses, memory becomes more defective. The most recent happenings are forgotten; for instance, the patient does not know how much time he spent in the physician's office or forgets the object of his visit. In the beginning only recent events are forgotten, the happenings of long ago being still retained; but later on the memory of events long past also suffers. Thus, dates of marriages, births of children, and important occurrences generally, are obliterated from the patient's memory. Toward the end of the disease the parietic cannot even recall his own name, or the names of his parents and children.

Impairment of Ideas.—The latest and least organized ideas are usually the first to disappear, while the associations most in use persist longest, and special aptitudes are likely to continue undisturbed for some time. For instance, physicians in an advanced stage of paresis have been known to write prescriptions faultlessly while failing in every other mental effort. In the early stages, intellectual loss is only partial, whereas in the later phases of the disease patients lose practically all their mental possessions. As the memory fails, its place in the intellectual life is often taken by the imagination. Numerous dream-like fabrications are recited; in fact, any fanciful thought which may accidentally enter the mind is given out as fact. Ideas can be suggested to the patient at will, and he may be easily argued out of them, for nothing appears fixed or stable.

Defects in Judgment.—In the early stages the patient's conversation reveals uncertainties, contradictions, and lapses, and as the disease progresses, he is easily disturbed by sound associations. The loss of judgment is also manifested by absurd business transactions and errors in professional work. Obstacles are disregarded and wild schemes are promulgated. The patients seem to disregard the interests of the en-

environment and their thoughts seem to circle about themselves. They eventually come to live in dreamland and everything depends upon their own ideas and wishes. So vivid are their "day dreams" that it is difficult to determine which of their expressions belong to the dream world and which are part of reality.

Early in the disease the patients may have what is called "insight" into their mental troubles, complaining of poor memory, difficulty of concentration and irritability. As time goes on, all judgment disappears and the patient seems to exult in his sense of well-being, becoming euphoric.

Hallucinations are not common in general paresis, but exceptionally they may constitute a prominent symptom.

Delusions.—The delusional ideas of the parietic vary considerably in different cases. They are usually of the fantastic kind, especially in the agitated and excited types. The patient refuses to think in terms of hundreds and thousands, but prefers to figure in billions, quadrillions, etc.; he fancies himself the king of kings, the god of gods; he contemplates himself even as the Deity itself; he possesses a million wives, has a billion children, is the creator of the universe. The delusions are shifting and variable; in fact, fixity of delusions speaks against the existence of general paresis. The delusions in paresis are always changing and often contradictory in the same moment; numerous ones come, others go, and some return. Delusions may even be suggested to the patient and may be readily changed at the will of the examiner. It is remarkable that the patient himself is usually unaware of his inconsistencies; for instance, the owner of the world may ask for a snuff of tobacco and complain of ill-treatment by his attendants almost in the same breath in which he announces his greatness.

Changes in the Emotions.—Changes in emotion are common. The patient becomes irritable and changeable. Joy alternates with sorrow, kindness with anger, discouragement is quickly followed by unreasonable optimism. In other cases the patient appears depressed, as in true melancholia with its suicidal thoughts and acts. At first he may have insight into his own condition and be deeply concerned in contemplating the approach of a serious disease. His mind may be filled with dark presentiments for the future—a state resembling true melancholia and often mistaken for it. As time goes on, however, it is soon noticed that the depression is only superficial and lacks the emotional background of true melancholia or manic-depressive insanity.

Changes in Character.—Patients who were formerly considered self-willed and of firm decision become vacillating and show instability alternating with obstinacy and perverseness. A patient with great initiative in his normal life may become a plaything in the hands of sharks. One who never drank or smoked may easily be seduced into a debauch of alcoholism or tobacco. Such patients are extremely suggestible; a discouraging conversation may lead to suicide. A knowledge that the owner of a certain article may have no immediate use for it will suggest to the parietic that he must possess himself of it at once.

And for a similar reason the sight of a beautiful woman may suggest rape to the parietic. Crimes are committed by him without a thought entering his mind that such acts are unethical and punishable. Convention and morality have no restraining influence upon his sexual longings. He delights in telling lewd stories, and consorts with people of inferior station; he holds in high esteem women of the demimonde and sexual perverts. Patients have brought to their own homes women of the street whom they introduced as their sweethearts, asking the consort of a lifetime to give up the best room in the house to such creatures.

PHYSICAL SYMPTOMS.—The physical signs of the disease are almost as numerous as the mental. They may appear either before the mental symptoms or not until dementia has become well advanced.

Headache.—One of the important complaints is headache, which may appear early, and is described by the patient as if the head were being held in a vise. There are also muscular weakness, shifting pains, pressure headaches, variously described as dizziness or emptiness of the head. These symptoms are so like those of neurasthenia that they have often been mistaken for that disorder.

Paralytic Attacks.—Of the motor symptoms, paralytic attacks—either epileptiform or apoplectiform—are important, as they occur in nearly one-half of all cases of general paresis.

Epileptic Seizures.—Epileptiform convulsions occurring in an individual for the first time after thirty years should always arouse the suspicion of general paresis. Not infrequently a jacksonian or a generalized epileptiform convulsion ushers in the disease. For years previous to the development of any other symptoms, convulsions have been known to occur. With the appearance of convulsions there is weakness and paralysis of the parts convulsed, the paralysis lasting for hours or days. After a convulsion the patient is usually left with reduced mentality, each attack subtracting something from the patient's intelligence. There are few exceptions to this rule. The convulsion itself does not differ essentially from that occurring in true or idiopathic epilepsy. There may be the same biting of the tongue, frothing at the mouth, the drowsy state following the attack; even the psychic equivalents of epilepsy are encountered.

Apoplectiform Attacks.—During these attacks the patients lose consciousness and appear to suffer from generalized attacks of apoplexy, which may be followed by monoplegia, hemiplegia and paraplegia. A distinguishing feature of the apoplectiform attack of general paresis as compared with true apoplexy is the observation that a general parietic may recover from his paralysis within a few hours or a few days. The entire attack may consist of loss of speech—aphasia, either complete or incomplete—and, while possessing all the characteristics of aphasia, is of exceedingly short duration, passing off perhaps after a few hours.

Reflexes.—The superficial reflexes are usually preserved, but the tendon or deep reflexes, especially the knee and Achilles jerks, may show anomalies. Knee jerks may be normal, exaggerated or unequal, reduced

lost. In a large number of cases the deep reflexes are reduced, showing posterior cord involvement. In the cases with increased deep reflexes there may be found other symptoms of pyramidal tract involvement—Babinski, Oppenheim, Gordon, and Chaddock signs. In addition to reduced or lost tendon reflexes, there may be other signs of involvement of the posterior columns, such as disturbance of the sense of position and of the deep sensibility. Likewise, ataxia of station and of gait, as well as anesthesia and lancinating pains of the tabetic variety are encountered. Cases in which the last group of symptoms occur have been termed taboparesis, or paresis of the tabetic variety.

Incoördination.—This varies from the slightest to the most intense degrees of ataxia. The ataxia of gait may be slight or severe, as in tabes. The slighter grades of incoördination are manifested in the handwriting and in the ataxic disorders of speech.

Tremors.—Tremor in the tongue, as well as in hands and fingers, may be marked or ill-defined. In most cases it can easily be brought out by having the patient perform voluntary movements, as protruding the tongue, extending the fingers or bringing a finger-tip in contact with the nose. These maneuvers usually result in a coarse, irregular tremor not unlike that of multiple sclerosis.

Disorders of Speech.—At first certain letters or syllables may be swallowed or slurred; the consonants “r” and “l” are particularly involved. This may be easily demonstrated by the use of such tests as “Methodist Episcopal,” “Truly rural,” etc. A favorite method is to have the patient repeat the following sentences: “Peter Piper picked a peck of pickled peppers”; “Around a rugged rock the ragged rascal ran”; “National Hospital for paralyzed and epileptics”; or, the rather difficult sentence, “Constitutional idiosyncrasy of the National Commonwealth of Massachusetts.” The paretic is rarely able to repeat these sentences and his mistakes often consist of a duplication or omission of certain syllables. A favorite test is to ask the patient to repeat one or two of these paradigmata, then request him to cite from memory one of the sentences. Usually the greater number of them are lost or a part of the sentence has completely dropped out of his memory.

Eye Symptoms.—In a large number of cases the pupils differ in size. In addition, Argyll Robertson pupil has been observed either in one or both eyes in different degrees—a fact which distinguishes it somewhat from tabes, in which the Argyll Robertson pupil is usually found bilaterally, more nearly of the same degree and more complete. In a small number of cases there is optic nerve atrophy, which is usually not of the same intensity on both sides. Less commonly the ocular muscles become paralyzed, causing either strabismus or ptosis of the persisting kind. This is usually an early phenomenon and belongs to the poploctiform attacks.

The physical and mental symptoms just enumerated represent in a general way the clinical picture of general paresis. It is now necessary to describe the several types of the disease, each of which, from the onset, runs a somewhat different course.

1. **THE DEMENTED FORM OF PARESIS.**—This is the most common type, which usually leads to simple deterioration without delusions and hallucinations. Remissions are rare and the course is rapid. While formerly the type of general paresis with grandiose ideas was considered the classical form of the disease, at present the simple demented type may claim that distinction. The symptoms may be best arranged under three stages.

First Stage.—The *onset* is gradual and may at first resemble a case of neurasthenia. Patients complain of inability to apply themselves at work; they lack concentration, and suffer from mental fatigue, pressure headaches and indefinite pains. The patient becomes depressed, irritable, violent, excited without cause, perhaps doing himself bodily harm. There may be an utter disregard of social conventions and neglect of the ordinary rules of esthetics and morality; he may appear in society with disordered dress, or with dirty hands or face and unkempt hair. An individual, formerly of irreproachable conduct, may, contrary to his former habits, begin to treat women indecently both in act and speech; he may frequent saloons and questionable resorts. Thus early in the disease patients are often brought into conflict with the law, for their tendency is toward immoral acts.

Meanwhile there is depression, anxiety and a feeling of physical illness. Sleep is usually disturbed, although in some instances the reverse is observed—the patient has a tendency to sleep more or less constantly. Some of these changes are often noticed by the people of his environment, who seldom recognize them as the beginnings of a serious mental disease but merely as the expression of immorality. During the early stages patients continue to work at their respective occupations; but it is readily noticed that, because of defective intelligence, they are unable to cope with difficulties arising in their work. There is, in addition, a remarkable lack of initiative; nothing new is undertaken. For obvious reasons, when the patient nevertheless attempts something new, he makes a complete failure of it.

The patients are usually good-natured, tractable, easily influenced by their comrades and often drink to excess. Some become very obstinate and self-willed. Women may neglect their household duties, forgetting to prepare meals for the family, or making them unpalatable, when formerly they were exemplary housekeepers and fine cooks. Soon consciousness becomes clouded and patients fail to comprehend their environment; they become disoriented as to time, place, and persons. They may become confused in their own home and fail to recognize friends and relatives. Transiently, a patient may become aware of his defects of memory, especially when his shortcomings are pointed out to him. In the early stages some cases have a foreboding of the oncoming softening of the brain and talk of committing suicide; sometimes they actually destroy themselves. There may be some anxiety with weeping and praying, and also increased irritability, some sexual excitement, aggressiveness and assaults. In general, however, the characteristic emo-

tional quality is that of progressive deterioration of the feelings. Gradually the patients become dull and apathetic. They are easily contented, provided the simplest needs of life are satisfied; give them ample food, drink and tobacco, and they have no complaint to make. They may be very polite, greet strangers with a smile and speak of their institution life in terms of high praise. At first there may be some insight into their condition; they may bewail their fate for having a poor memory and slowness of thought, but increasing deterioration gradually dulls the faculty of self-criticism and produces a sense of well-being and perfect confidence in their own abilities. While the majority of the patients suffering from the dementing type of paresis are readily suggestible, there are some who are inaccessible, repulsive and surly, answering questions as if angry, and refusing to comply with any request made in friendly terms.

Paralytic attacks occur in almost one-half of the cases.

Physical signs are either absent or inconspicuous during the *first stage* of the disease. The majority of cases are diagnosed as hysteria by the general practitioner, and because of the absence of neurological signs at this time, even psychiatrists have been unable to make a definite diagnosis without calling the laboratory to their assistance.

Second Stage.—The mental weakness is well defined; memory fails and the patient shows indifference to his surroundings. The parietic speech becomes more noticeable. There occur paralytic attacks, which are sometimes very mild and characterized by dizziness or transient losses of consciousness, following which there is a further dulling or weakening of the mental powers. The neurological symptoms are either unchanged or they become more marked; especially is there a tendency for ataxia to increase. On account of intellectual weakness, the patient is incapable of mental labor and, because of the weakness or ataxia of the extremities, he is also unfit for bodily exertion.

Third Stage.—The mental weakness sinks to dementia; memory has almost completely disappeared. The simplest questions are no longer answered—the patient becomes wholly indifferent. The muscular paralysis becomes more complete and an attendant becomes indispensable; urine and feces pass involuntarily. Apoplectic attacks followed by paralysis increase the patient's helplessness. He may either die in such an attack, or aspiration pneumonia or other intercurrent diseases may carry him off.

During the course of the dementing type of general paresis there appear occasionally evidences of depressive or maniacal phases, which may exist in the patient along with the other symptoms. The type of the disease is not thereby altered, for delusions of grandeur and of micromania are only brought out by direct questioning concerning them.

In order to illustrate the dementing type of general paresis, the following condensed report of a clinical demonstration of three cases at different stages of the disease is here given:

CASE XIII.—The patient is 35 years of age and an actor. His speech may be termed clumsy, stammering, indistinct, with a tendency to slur over syllables and words. The syntax is bad, there is a waste of words: too many verbs and adjectives and too few nouns are used. His fifteen minutes' conversation can be easily condensed into a half-minute talk. In conversing with him it becomes apparent that he still has insight into his condition—he knows there is something wrong but does not know what. He informs us that he was working until recently as a member of an acting team, doing dialogue acts and some dancing. Of late he could not keep up and was compelled to engage another actor to take his place, because he would forget his part and his speech became hesitating. From an actor we certainly expect more than the patient can furnish. It is well known that in this disease a patient may go on with his routine work for a much longer time than with work to which he is unaccustomed. This man has been in the habit of talking rapidly. He now drags out his words and sentences, which indicates quite a serious defect. If, for instance, a bookkeeper whose work has been to add columns all his life should become unable to add up 125 and 79, you would infer that he is in a serious way, while in an ordinary laborer, unaccustomed to figures, this test would mean very little. Our patient, therefore, shows a rather grave symptom because of his marked speech defect. When asked to repeat the usual paradigmata, as "Peter Piper picked a peck of pickled peppers," etc., he hesitates and refuses to follow his examiner, because he claims his speech is poor on account of the removal of his tonsils six months ago. This "fishing" for excuses is quite common in a beginning paresis when the patient is still conscious of his defects. When questioned as to dates and persons he seems to be well oriented, also as to the environment.

The physical signs are speech disturbances, tremor in face and tongue, also slightly in the extended hands. The knee jerk is exaggerated on the left, but normal on the right side; Achilles reflexes are normal. The pupils show a difference in size and are not quite circular in outline, but there is no Argyll Robertson phenomenon. There is no incoördination, either of station or of gait. There is a distinct history of syphilis of 20 years ago and the Wassermann findings are positive on blood and spinal fluid.

The mental and physical symptoms in this case are few and ill defined; there are no definite physical signs beyond the speech disturbance and the irregularity of reflexes, tremor in tongue and facial muscles, but these are sufficient for a diagnosis.

This patient suffers from an early stage of simple dementing general paresis.

CASE XIV.—The patient is 37 years of age, an American, who entered the hospital in a state of confusion. From his gestures and conduct he appeared to be suffering from visual hallucinations, but he was non-communicative; the history was obtained from relatives.

Ten months ago, while at his work, he suffered from a "stroke," which caused him to fall and become paralyzed and aphasic. After a day or two recovery took place: he could speak and work as usual. A few weeks later similar attacks occurred at intervals of one or two weeks. Each attack was either accompanied by spasms, vomiting and cramps, or a distinct paralysis of the extremities took place. The entire attack usually lasted a few hours and was regularly followed by recovery. During his stay in the hospital a number of such attacks were observed. When free from apoplectiform and epileptiform attacks he was generally useful about the hospital and everybody thought he was improving; just then another series of attacks would be noticed. His speech would return after the spells, but was hesitating and indistinct, until it became permanently incoherent and mumbling; the patient has now become weak and bedridden.

These quick recoveries from transient apoplectiform attacks are characteristic of general paresis. Usually, as in this case, dulling of the mental faculties, complete apathy and loss of memory are combined with the attacks. For instance, the patient did not know his name the day previous, but he knows it now and may again forget it in a few minutes. During this demonstration he seems improved. This is his bright day, for he answers correctly a number of questions as to age and onset of symptoms. His speech is typical of the slurring and indistinct paretic variety. The patient denies a history of syphilis, but there is no doubt that he suffered from the infection many years ago. Physical signs are abundant; the pupils are rigid to light, but respond in accommodation—are of the Argyll Robertson type. The tongue is extremely tremulous, there is a peculiar, irregular, pendulum-like to-and-fro movement. When asked to count from one to fifty aloud, the mere attempt to do so is sufficient to throw his facial musculature into a coarse, irregular, vibratory tremor, compelling him to stop at seven. There is a typical Romberg sign and the gait is extremely ataxic. The Wassermann is strongly positive on blood and spinal fluid; the Lange test shows a typical paretic curve.

This is a case of the simple dementing variety of general paresis with a rapid progress and apoplectiform and epileptiform attacks, without the violent mental episodes found in other subgroups of the disease.

CASE XV.—This case is somewhat more advanced and may be called terminal. Patient is unable to understand questions put to him, and consequently no history is obtainable. When he attempts to answer the simplest questions, his lips begin to quiver and tremble and the entire face is thrown into a continual vibration, causing complete failure. In an attempt to protrude his tongue upon command, he only succeeds in causing the face to become more tremulous. There is a constant coarse and irregular tremor in both hands and fingers, which becomes aggravated when he attempts to extend them. Not only is there tremor in the upper extremities, but the lower extremities are

also involved in a similar tremor when he attempts to walk. The mentality is impaired to such a degree that the simplest conversation is impossible; he appears bewildered and gropes for words and ideas. There are distinct Argyll Robertson pupils and the knee and Achilles reflexes are exaggerated. The Wassermann is four plus positive on blood and spinal fluid.

This is undoubtedly a terminal case of the dementing type of general paresis.

2. **EXPANSIVE FORM OF PARESIS.**—The characteristics of the expansive form are delusions of grandeur, a prolonged course, and greater prevalence of remissions.

The onset is usually slow. There is noticed increasing difficulty in mental concentration, failing memory and poor judgment; in fact, a radical change in character, and with it increased irritability. In addition the usual physical signs are observed, as fainting spells, transient speech disturbances, attacks of syncope and headaches.

Resembling certain types of manic-depressive insanity, patients may at first pass through a stage of depression, with self-accusations, delusions of persecution and states of anxiety, followed by excitement and exaltation. In many of these cases excitement, with elation and grandiose delusions, opens the scene, while states of depression are of short duration and appear only occasionally. In those cases in which despondency was the prevailing mood, this disappears either gradually or suddenly and a marked feeling of well-being takes its place. Patients are busy, affable, talkative, and constantly planning new schemes, are about to build a million dollar apartment, buy up all the oil wells and marble palaces, and, in short, change the aspect of the universe. At first the delusions of grandeur are still within reach of the imagination, having an air of possibility about them; later they exceed all bounds of probability and are as shifting as the clouds, are absurd in their content, and carry within them the evidences of a deranged mind. Added to delusions reaching the most unheard of heights is an extreme restlessness, so-called psychomotor activity, yielding the characteristic picture of megalomania. Asked how they feel, the answer is invariably: "I never felt better in all my life." To prove this assertion, they continue to flex their biceps, pointing to its firm contractions, beat their chests, and speak of their enormous physical strength. The abundance of strength is unimaginable; and "I can lick the world" is not an uncommon expression. The better educated boast of their beautiful oratorical style, fine syntax, fluency of foreign tongues, great knowledge of music and art. They count among their friends presidents and they are being consulted by the kings of Europe on matters in which they have become recognized experts. Their plans are quite utopian—poverty will exist no more; disease and war must be permanently banished from this globe, and they have the means to accomplish this feat. Their business is better than ever; their profits this year will amount to five billion dollars. The lowliest station in life is not incompatible

with the most wonderful achievements; a day laborer has suddenly invented an airship which will carry passengers to Europe in two hours. They usually speak in figures of millions and billions, despising small sums of money and small things in life. They appear mostly in a happy mood and develop a sense of well-being—euphoria—entirely at variance with the true state of affairs.

Absurd delusions, multiplied in the most diversified ways, are merely a sign of progressive mental weakness. In women the tendency to expansive delusions is not as well marked; their great ideas center mostly about little things connected with their household duties; they bake the finest cakes, prepare the most wonderful meals and can raise the largest families.

In this type of general paresis consciousness is somewhat clouded—patients are too much absorbed in their grandiose delusions to acquaint themselves with their surroundings. In consequence there is disorientation for time and place and persons. They are mostly occupied with self and the many delusions, and rarely know the month, day, or year. The invariable excuse is, "I was too busy to read the newspapers, consequently I did not look at the date." At first, thoughts may be coherent, although during great psychomotor activity there may be incoherency, distractibility and flight of ideas.

The *emotional attitude* usually corresponds to the content of the delusions; the patients are cheerful, hopeful and exalted. Everything pleases them. The fellow patients in the hospital are the finest neighbors in the world; the coarse hospital fare is as good as that of the finest hotel; the doctors are the politest people in the world. Mingled with the most hopeful emotions there occur short periods of depression, when patients cry bitter tears, being almost unconsolable, like little children. The proper suggestion by physician or attendant is usually sufficient to reestablish the old euphoric mood. As a rule these patients are extremely suggestible, are easily led from one subject to another; only occasionally do they show anger when somebody disagrees with them. Later in the course of the disease the patients become uniformly satisfied and pleased with the most trifling attention, even when they are bedridden and suffering from terminal paralysis and contractures.

In many cases psychomotor activity reaches a stage resembling the manic phase of manic-depressive insanity. Patients are active all day and remain awake nights, constantly doing something,—talking, writing, traveling about from place to place, always working on the most improbable schemes. This leads them to extremes: they may become reckless, aggressive and impulsive. They may be possessed of anxiety and senseless fears, which cause them to cry out from fright. The periods of great excitement usually last only a few hours and leave the patient in a state of deterioration.

In conduct and act the patient becomes foolish, lacks judgment and becomes indifferent to his normal standard of morals. He may swear, smoke, drink, and indulge in extramarital normal and abnormal

sex practices; he may commit thefts and assaults, but in all of his misdemeanors there seems to be an absence of plan and disregard of any but his own interests. In the further progress of the disease, great activity ceases, patients become quiet and appear contented, always "feeling fine." Remissions of practically all symptoms occur in one-third of the cases. The expansive form may pass over into the depressive, or *vice versa*, when the disease picture simulates true manic-depressive insanity.

The following is a case of the expansive form of general paresis:

CASE XVI.—The patient, a young tailor, 34 years of age, appears very happy, pleased with himself and his environment. He states that he is not married, but can have millions of girls and married women. He wants to get out and work, for he can make a million dollars a week. He knows he feels fine and never had a better day in his life. The reason for his stay in this place is merely to find out how the other half of the world is doing. In his opinion the treatment here is excellent and he has a good notion that he would rather stay here than elsewhere.

Physically, there is a tremulous tongue, very marked, coarse tremor in both hands. The right pupil is larger than the left. All the tendon reflexes are exaggerated, but there are no pathological reflexes. There is a beginning Romberg sign and the face is asymmetrical.

This is the usual type of exalted general paresis.

3. AGITATED FORM OF PARESIS.—Characteristic of this form is a degree of psychomotor excitement more marked than in the expansive form. The onset is rather sudden and accompanied by numerous delusions and great clouding of consciousness. The entire course is of short duration. Prodromal symptoms, observed in the other types of general paresis, are usually absent; there is rapid development of extreme megalomania. The patient suddenly becomes energetic and overflowing with good health. He is possessed of untold riches, knowledge, beauty and wisdom. Indeed, so powerful has he become, that he has even created God and the universe, and considers himself the final super-god. At a mere command by the patient worlds may spring into existence and others may perish. Not satisfied with knowing themselves to be all-powerful, they wish to coerce the rest of the world to "sit up and take notice" of them. They may even order war and command their imaginary generals to appear at the war council. Some have great ideas as to economic reforms, which must be most sweeping and radical, and stop at nothing, at least as far as they are concerned. One patient has made special arrangements to live eternally and wishes to regularly entertain God and the entire heavenly crew, another has a longing to befriend Satan, who lost his seat in heaven, which is going to be restored to him shortly. One patient's house must be built of the purest gold and finished within three days. Another must be consulted for advice on state matters by rulers and lawmakers, for he is

the most renowned statesman living. There is practically no limitation to the parietic's powers. Meanwhile they are talking, shouting, stamping with their feet, and always busy doing something.

While the mood is generally elated, there are attacks of the most profound depression and hypochondriasis, when everything seems destined to destruction. These last attacks are usually of extremely short duration and are quickly followed by shouting, singing, dancing and talking. Constantly active, they shift quickly from one thing to another, never retaining long a single idea. Patients are too busy to attend to their bodily wants; they forget to eat, move their bowels, or bathe the skin. Others eat several times in succession, forgetting that they had just finished their meal. Hallucinations of sight and hearing are occasionally observed, but these are not part of the clinical picture. There is flight of ideas—incoherent thought. The emotions are also changeable; they may be good-natured one minute and very irritable the next, especially toward those who doubt their greatness or oppose them in the slightest degree. Patients of the agitated variety lose weight and the temperature may become subnormal. In about one-fourth of the cases there are remissions.

Some cases of the agitated form present the most extreme degree of excitement and clouding of consciousness, and terminate fatally within a few weeks. These cases have been termed *galloping paresis*. The patients are completely confused, unable to comprehend anything and do not answer questions. They are noisy—shout, cry, sing and dance almost at the same time. More or less constantly in motion, they refuse to eat and do not sleep, exhausting themselves perceptibly day by day. Epileptiform and apoplectiform attacks may “break” the monotony or the patient. After a short period of the restless phase of this condition, patients sink into stupor, during which the movements become uncertain and tremulous. The end usually comes by infection, pneumonia, or inanition.

The following private case illustrates well the agitated type:

CASE XVII.—The patient is 32 years of age, an unmarried Greek, who subsequently admitted having had syphilis 15 years before the beginning of his present illness.

He came under the writer's observation about 20 months ago while under restraint in a fashionable sanatorium. Delusions of grandeur were in abundance: the patient was an intimate of the President of the United States, of the Mayor and the Governor. While he was enumerating his many distinguished acquaintances, there was a sudden interruption caused by a noise in the corridor. Immediately he screamed at the top of his voice: “Come in, Mr. President, you are expected.” At other times, he would answer a knock at the door by saying, “Come in, Mr. Mayor,” and frequently he remarked that he expected a telephone call from some distinguished person. This occurred during the Great War and when some Greeks residing in this country, the wife of whose former king was a Hohenzollern, continued to sympathize with Ger-

many. The patient had expected the mayor of Chicago, whose sympathies were also believed to be with the Germans; he was, therefore, extremely impatient with the writer for visiting him at this time. He refused to answer any interrogatories because of lack of time, having a previous appointment with the Mayor, who was coming to consult him on the European question. He was too busy with peace problems and he needed all his time and energy for that purpose. Besides, he never felt so well, and saw no need of wasting the writer's time and his. He asked the former to make way for the Mayor—who never appeared. When visited again, he was very talkative and discussed the War, which, he was convinced, he could "settle" in five minutes. He was expecting hourly messages from President Wilson, who was about to conclude peace with Germany, though he thought the people of America were eager to continue the fight. Being a personal friend of the President, the patient was anxious to help him in every way. At this time he became extremely noisy and his speech became incoherent and rambling. As during subsequent visits, the writer and his patient became friends; he began to shower millions of dollars upon the writer, with directions to buy a \$10,000 automobile at his expense, and requested the favor of permitting the writer's wife to accept of him a diamond brooch valued at half a million dollars. He mentioned fabulous sums of money of which he was the possessor, and he was positively determined to make mankind happy with his prodigious wealth. In addition, he made it clear that his powers of persuasion far exceeded those of William Jennings Bryan; that he, the patient, was certainly a better orator, and intended to address an audience of 75,000 persons—the largest gathering of people the world had ever witnessed. He thought his voice was adequate, but needed training, because he feared it was not quite far-reaching. It, therefore, became necessary to begin a course of voice training for the occasion. On the following day the patient was hoarse and almost exhausted. The nurses reported that he had rehearsed his speech all night, for he was anxious that every man, woman, or child of the 75,000 people who would come to hear him should catch the meaning of his every word. He begged the writer to promise that he and his family will occupy the platform when he shall deliver his great oration. Peace to him was assured, provided he could deliver the great speech. From that time and during the next few days, he rehearsed his speech day and night. The consequence of these rehearsals can be imagined: for blocks around the people would stop on the street, attempting to locate the evildoer. Once they discovered his location, it was impossible for the patient to remain. He was summarily dismissed, and without ceremony, from four different "rest cures," because of the intolerable noise, which disturbed the peace of the vicinity, while the patient was laboring toward peace of the world. The delusions of grandeur and the violent moods persisted during a period of eight weeks. Gradually the patient became more docile, permitting radical treatment to be administered. A short remission occurred, which lasted three weeks, during which time the

patient remembered in part the happenings during the exalted phases of his disease. Then an exacerbation of symptoms occurred; this was only of short duration. After an extensive course of Swift-Ellis and mercury treatment, the patient cleared up; in other words, a remission occurred lasting to the date of this writing. He has now a clear conception of our past and present relations with the enemy, and knows that his sympathies had been misplaced. His brothers assured the author that his mentality shows no deterioration, though he spoke to the writer recently of the "tragedy of life" without emotion.

Mentally the patient continued to improve, but physically there developed marked ataxia and Argyll-Robertson pupils. The Achilles reflexes, previously present, have disappeared, and the patellar reflexes have become unequal—the right showing a faint reflex, while the left is entirely absent. In addition, there have developed sharp lancinating pains, and the entire picture has become one of tabes minus general paresis. Though the spinal fluid repeatedly showed a four plus positive finding, the reaction had become negative during the remission. At the end of six months there was a two plus positive Wassermann on the spinal fluid with a negative blood reaction. Finally the blood and spinal fluid became again negative and the patient returned to Greece. It may be of interest to record that the writer's sworn statement saved the patient from being sent to the front, for he had been put into a uniform soon after his arrival in Greece. From his relatives the writer learns that he continues well.

4. DEPRESSED FORM OF PARESIS.—Despondency and depressive delusions are the dominant characteristics of this form of general paresis. Patients are usually introspective from the beginning; they feel themselves becoming inefficient, tired and weak. They suffer from the numerous pains and aches of the neurasthenic—pressure headaches and the various backaches, loss of sexual power and of sluggish bowel action. They constantly worry and become extremely hypochondriacal, claiming they are doomed, as no cure is possible for them. One physician after another is consulted, but each one in turn is told by the patient that there is no use wasting time on him, for he knows that his case is the worst in existence. It is because of these early symptoms that patients are considered neurasthenic, hypochondriacal or hysterical. Another diagnosis frequently made at this time is that of melancholia.

At first complaints have an adequate emotional background; later they are senseless. Believing that the stomach is made of glass, they refuse to eat; besides, digestion has ceased, the bowels have not moved in months, and the tongue consists of a mass of corruption. The throat is clogged by a carcas, there is no taste or smell left, and the vital fluids are escaping in the urine. Some of these patients may be found sitting for hours without the slightest attempt being made to change their position. The reason assigned may be fear of instant death.

In many instances the hypochondriacal state is accompanied by delusions of sin. They accuse themselves of all sorts of crimes, from hav-

ing spoken ill of a friend to having committed the most atrocious murder. Of course, they ask for no sympathy or pity—to be burned at the stake is what they deserve and what they confidently expect. They have enjoyed at the expense of people's suffering; they have always lived a lie and must now get their punishment. Occasionally patient interrupt the conversation by an anxious look toward the door, asking "When are they coming to take me to the scaffold?"

There are not only the delusions of sin, but also those of persecution, accompanied perhaps by visual and auditory hallucinations. Remarks against their person or their family are overheard; conspiracies are being hatched against everybody dear to them. Whole armies are being mobilized against their acquaintances. Occasionally they see their enemies entering the room, standing before them with drawn swords, and threatening death.

The consciousness becomes confused and illusions may occur. Time, place and persons are mistaken, but one thing seems certain to them: everything has direct reference to themselves. This type of depression may aptly be compared with agitated melancholia or hypochondriasis, which conditions are almost indistinguishable from this form of general paresis. The patient, like the true melancholic, paces the floor, wringing his hands and begging for death. "Kill me, doctor!" is a favorite expression with them. In this disease, however, there is always present a note of the nonsensical and ridiculous, with an air of insincerity about their complaints. The condition continues to grow worse, and finally the patient remains in bed huddled in a corner, awaiting the end. Others wish to hasten the end by attempts at suicide, which are made in such a whimsical and ineffectual manner that they only succeed in mutilating themselves. The periods of actual anxiety are of short duration, thus again differing from true depression; they usually alternate with longer periods of complete apathy, or even a period of spurious exaltation may supervene.

States of stupor may appear, during which patients become mute, being oblivious to their surroundings, refusing food, and being negativistic in every way. The resemblance of dementia præcox to catatonia is striking; there are the muscular rigidity, negativism and mutism. The stuporous phase may persist for weeks or may appear on alternate days, with approximately rational days between. The depressed form of general paresis constitutes about one-fourth of the cases and occurs in persons past 40 years of age. Remissions may be expected in about 10 per cent. of cases and paralytic attacks in 25 per cent.

The following case is fairly typical of this form of general paresis.

CASE XVIII.—The patient was a man 55 years of age, but rather senile looking. At the time of the first examination he was extremely emaciated, though not bedridden. When anybody came into the ward the patient rushed forward, complaining of his treatment and bewailing his fate. His favorite method was to protrude his tongue, touching it with his fingers, and then to begin a most graphic description of how his tongue was rotting away and how physicians and nurses combined to cause his death. At the same time he felt sure that nothing

could be done for him, and that he must be permitted to starve himself. He refused to take food not only because of his tongue, but also because his stomach had been transformed into glass, which prevents the secretion of gastric juice.

Physical examination revealed Argyll Robertson pupils, exaggerated reflexes in knee and Achilles tendons, tremor in tongue and hands, and a slight degree of ataxia of station. The patient ultimately became bedridden and died from an intercurrent pneumonia.

The prevailing features in this case were hypochondriasis and melancholia of a rather constant type, but with the most absurd somatic delusions.

5. **TABOPARESIS.**—Cases occur in which to the mental symptoms are added many of the classical signs of tabes, such as the loss of the tendon reflexes, Argyll Robertson pupils, lancinating pains, ataxia of upper and lower extremities, Romberg sign, hypotonia, crises and arthropathies. Some neurologists regard tabes as a spinal paresis and paresis as cerebral tabes. The writer's experience bears out the statement made by others, that taboparesis is not exactly the same as tabes and general paresis, but that the disease is a composite, showing differences in either component. In a general way the tabes is ill-defined, lacking many of the usual symptoms, and the mental symptoms of paresis are somewhat milder.

The following case is fairly representative of the group.

CASE XIX.—Patient is a widow, 41 years of age, a waitress. She entered the hospital because of pain in the epigastrium, double vision, pains in the feet, severe headaches, and loss of memory. About three years ago she began to see double; there was no other complaint at that time. Recently she began to suffer from severe headaches and pains in the feet. She is the mother of two living children; one child died as an infant from summer complaint. An examination reveals a ptosis of the left eyelid and there is a left internal rectus paralysis, causing diplopia when patient looks to the right. The right pupil is larger than the left. There is complete Argyll Robertson pupil in the right eye and sluggish light reflex in the left. The knee jerks and Achilles reflexes are absent.

Mental tests reveal the patient's inability to name the day of the month or year correctly, and, besides, she mistakes the writer for some one else. The speech shows marked hesitancy and there is elision of syllables when she repeats the phrase: "Around the rugged rock the ragged rascal ran." The tongue is not tremulous and no tremor can be seen in the extended hands and fingers. There is no paralysis or asymmetry of the face.

Diagnosis is based on the mental involvement and the physical findings of sluggish pupillary reflexes, absent tendon reflexes and speech disturbances. The Wassermann test is strongly positive and the Lange colloidal gold test is classical, the first four tubes of the series being completely discolored. This is the typical paretic curve

The patient lives in the past, remembers nothing and feels happy, regardless of her environment.

In this case we have tabes symptoms combined with those of paresis—in consequence it is diagnosed as taboparesis.

6. JUVENILE PARESIS.—This form of general paresis is not as rare as formerly believed. Many of the cases have been diagnosed as idiocy or as dementia præcox. Laboratory and careful clinical examinations should insure a correct diagnosis in every case. Every youthful case of supposed mental backwardness is not dementia præcox or idiocy; and it is well to think of the variety of paresis called juvenile paresis.

Paretic children have either been normal or brilliant at school. Then suddenly, as it were, the child's mentality becomes arrested. Poor memory and gradual dementia appear, with or without the episodes described as characteristic for paresis. Childishness, exaltation and depression, fears and anxieties are common symptoms. Epileptiform and apoplectiform attacks may appear as in the adult. The age of onset is from 7-15 years after infection, the same as in the adult types. The histopathological changes in the brain do not vary essentially among the different forms of the disease. A much better idea of the disease can be gained from a perusal of the next three case histories than from any amount of theoretical description.

CASE XX.—Patient, a young man aged 20, was brought to the writer's clinic at Northwestern University Medical School for poor memory, nervousness, trembling and impaired speech. Quite interesting is the fact that the family history throws little light on the case. Patient's mother, 51 years of age, had been married at 19, had never been sick, and comes from a healthy family; her mother is well at 80, father died at 86, three brothers and one sister are well. The patient's father is 54, and always had good health; his father reached old age, but the mother died young.

The obstetric history of the patient's mother is as follows: The first two pregnancies resulted in abortions of two and three months respectively; nineteen months after her marriage a son was born, who is now 30 years of age and in good health; then came another son, who is healthy; and two years later, a daughter, who is 25 years old and well. Henceforth the offspring showed little vitality. A daughter died at four weeks, because of difficulty in nursing; the next child, a boy, lived two hours; then our patient arrived.

He was born in normal labor and seemed well up to the age of two months. Then appeared an eruption on the face; the lips and mouth became ulcerated. His nose was inflamed and there was difficulty in breathing and nursing; he was extremely restless and cried most of the time. At this time also he suffered from a severe eye disease, which was cured in one week. There were no convulsions nor headaches. After six months' treatment the patient completely recovered and remained well until the beginning of the present trouble. He walked at nine

months, talked at eighteen, and was always considered bright, being a fairly good scholar. The general health continued good until the beginning of the present symptoms. His habits were generally good, though he was a moderate smoker and drinker.

About one and one-half years ago, before consulting the writer, his character gradually changed. He became irritable, peevish and would occasionally leave the house for a few days without an assignable cause. The parents ascribed these irregularities to bad temper, until they discovered that his physical health began to deteriorate. Then they thought of drug addiction, which was a mistake. A little less than a year ago his moodiness changed into euphoria; he was usually happy, laughed over trifles, developed an enormous appetite and thirst and, in contrast to his former self, he became very tractable.

While riding in a street car six months later he experienced twitchings in his face accompanied by loss of speech, lasting for about an hour. This was followed by occasional attacks of transient loss of power in arm and leg, which always terminated in complete recovery. Last winter while dancing—being a vaudeville dancer—he became dizzy and fell off the stage. Since then his condition has been steadily getting worse; speech had become indistinct and continues to grow more so. For the past week on alternate days he has been mute, inactive, disinclined to eat, but obeys commands when repeatedly urged. On other days he is talkative, reads the newspapers, turns to the parents for information, which the writer tries to elicit from him.

Examination.—In appearance the patient is rather boyish for his age, looks somewhat frightened and has a peculiar vacant stare. When he attempts to speak, the lower facial musculature is thrown into a state of trembling. The pupils are large, slightly irregular, do not respond to light and but little in accommodation. Immediately upon protrusion of the tongue a coarse, irregular to-and-fro tremor is seen, which it shares with the musculature of the mouth. The speech shows the typical parietic stumbling, trembling and slurring utterance. The patient is oriented as to time, place and persons.

There are no delusions of grandeur and there is a fair degree of insight into the condition; he is conscious of memory defects. Mathematical problems requiring but small feats of memory are beyond his reach.

An examination of the eye-grounds reveals normal findings bilaterally. There are white lines radiating from each corner of the mouth, indicating cicatrices of former lues. The teeth are not of the Hutchinson variety, but the cutting edges of the upper incisors are rough and worn away.

The reflexes are brisk in the upper extremities, the knee jerks about normal, but Achilles responses are absent. There are no disturbances of sensation, but there is a slight Romberg and some incoördination in the upper extremities. The viscera are normal.

Summary.—We have early eruption, snuffles, short period of ill health eventuating in recovery. During 18 years there is freedom from

all symptoms; no headaches, paralysis, or other evidence of lues. Then develop, rather slowly, signs of emotional and intellectual disintegration, transient attacks of motor losses, speech disturbances and tremors characteristic of general paresis. Of interest is the absence of a history of syphilis in the parents. However, the birth of two short-lived children preceding that of the patient, in conjunction with his early symptoms, and the scars about the mouth, enable us to make a diagnosis of congenital or early lues, followed by general paresis.

CASE XXI.—This case was admitted to Cook County Hospital for nervousness. The father is living and well, mother died of pleurisy at 38. There are three living brothers and one sister; several died in infancy.

The patient was well until the age of 9, when he developed suppurating glands on the right side of the neck, for which an operation was performed, leaving a large scar. He was a moderate drinker until five years ago, when he contracted gonorrhea and chancroid. Careful probing for the existence of secondaries yields negative findings. The patient is married. There were two pregnancies: the first one resulting in abortion, the second in a healthy child, now 18 months old.

About three months ago he had what appears to have been an attack of grip of moderate severity, but which left him somewhat nervous and caused him to enter the hospital. He states that his attack of grip is responsible for his weakness and nervousness. He complains of occasional dizziness and he is certain that for some time his memory has not been good.

Examination reveals a fairly well-nourished young man with a rather insipid looking asymmetrical face. Around the angles of the mouth several linear scars can be seen radiating in all directions. When he speaks, a slight tremor can be seen playing around the lips, which becomes more pronounced when he is angry. The pupils are irregular and do not respond to light and but slightly to accommodation. The fundi are normal. The eye muscles functionate correctly and no nystagmus is observed. Abdominal and cremasteric reflexes are normal; likewise the deep reflexes in the upper extremities. The knee and Achilles jerks are equally exaggerated; ankle clonus and Babinski's phenomenon are not elicitable.

Sensory disorders are absent. Coördination is normal in the upper extremities, but there is some ataxia of station and gait in the lower extremities. There is slight tremor in hands and fingers. The protruded tongue shows a jerky, coarse tremor which extends to the lips. Speech is hesitating and in a lengthy conversation becomes tremulous in character. The usual paradigmata can be repeated, but there is noticeable an omission of syllables and inability to pronounce the letter "r."

His arithmetic is exceedingly faulty: Examples: $9 \times 9 = 72$; $100 - 21 = 78$, etc. Memory tests show defects which are not very pronounced. There is complete orientation as to time, place and persons.

Summary.—We have a young man with Argyll Robertson pupils,

exaggerated reflexes, slight incoördination, coarse tremor in face and tongue and speech disturbance, with impairment of memory and inability to add and multiply simple sums. In addition, his emotions are extremely changeable: a word or look is sufficient to arouse his temper. There are enough somatic signs to make the diagnosis; and the mental signs, while not very pronounced, are characteristic of general paresis. The finding of the silvery white lines around the mouth is highly suggestive of infantile syphilis. The case, therefore, appears to the writer to belong to the group of juvenile paresis, preceded by congenital or early acquired lues.

CASE XXII.—The patient is a girl, 18 years of age. She was brought to the hospital for nervousness and speech difficulties. At 13 she entered the sixth grade and since then has done housework. An examination brings out the fact that her attention is lagging and general information and memory are poor. There are no illusions or hallucinations of any kind and orientation is good. Of course, the nearest thought is dementia præcox, but this diagnosis can only be made after a careful physical examination and laboratory tests. Because a patient is young, we have no right to diagnose every case as dementia præcox.

Examination.—Physical examination reveals unequal pupils which do not respond to light. The knee and Achilles reflexes are lost. With eyes closed and feet approximated, there is considerable blinking and twitching of the lids, but little or no swaying. The speech is indistinct; the paradigmata are not well repeated. The tongue shows an irregular, jerky tremor; likewise the hands. General intelligence and information are much impaired. Gait is somewhat unsteady, slightly ataxic. When asked how she feels, the answer is: "I have pains in the knees." She describes the pain as quick, like stabs which come and go and feel like needles and pins. There are no sphincter disturbances.

In examining the teeth, we find them deformed and one incisor looks like a typical Hutchinson's tooth. There are no evidences of scars around the corners of the mouth. With lost tendon reflexes, lancinating pains, the diagnosis taboparesis can be made. Wassermann is positive on blood and spinal fluid.

LABORATORY FINDINGS.—For a detailed discussion of laboratory findings the reader is referred to the same heading, at the beginning of this article (p. 105).

In general paresis the Wassermann test on blood and spinal fluid is usually strongly positive, the spinal fluid giving a positive reaction with the smallest quantities—0.05-0.2 c.c.—a point of differentiation from other types of neurosyphilis. An additional point is the fact that not only is the positive Wassermann seen in its greatest intensity in general paresis, but once present it is with difficulty changed into a negative one and often becomes "Wassermann fast." It shares this peculiarity with the most serious forms of neurosyphilis, especially those

which are about to pass into general paresis or when tabes is becoming taboparesis.

Kaplan, from his large experience in the laboratory, attempts a division of parietic serological findings into (1) the full-fledged case of general paresis which give positive Wassermann on blood and spinal fluid, as well as globulin reaction and lymphocytosis (17-50 cells); and (2) those cases in which the blood-serum, globulin and cell-count are positive, while the spinal fluid is negative for Wassermann; and (3) a small group in which the blood is negative, while spinal fluid, globulin and the lymphocyte count are positive.

As regards an increase of lymphocytes, or so-called pleocytosis, Kaplan thinks that a decided pleocytosis marks early paresis, while in the majority of clinically distinct cases of general paresis with slight pleocytosis and negative blood Wassermann reaction, he assumes an advanced process and one offering but little hope.

The same author, having seen negative Wassermann tests in a number of cases of general paresis, comes to the conclusion that there is only one uncontrovertible rule with reference to positive and negative Wassermann, which he formulates as follows: "A positive serology is not compatible with a nervous disease of non-luetic origin, while a negative serology may be obtained in a syphilitic nervous disease." In the writer's opinion, whenever a case clinically diagnosed as general paresis presents negative findings on blood and spinal fluid, the diagnosis of paresis should be reconsidered until such time as further examinations shall either prove or disprove the clinical diagnosis.

The Gold Chlorid Curve.—A definite reaction of the precipitation test by Lange has been observed in paresis and in cases of taboparesis. This consists in the complete discoloration of the first three or four tubes, with a stepladder curve in the remaining dilutions. This test now occupies a prominent place in the serology of general paresis. It is not characteristic of neurosyphilis in general, but may be considered a special test for general paresis. Tabes does not give it, neither does cerebrospinal syphilis. In some respects it is the most reliable laboratory test for general paresis, for when the Wassermann test is doubtful or absent, the Lange test may give a positive finding, while it is never positive in non-paretics.

Diagnosis.—**NEURASTHENIA.**—During the neurasthenic phase of general paresis the disease may be mistaken for true neurasthenia. There are the symptoms of fatigue, depression, pressure headache, lack of mental concentration, forgetfulness, and the numerous other complaints usually found in the great neurosis. The circumstance that the so-called neurasthenic does not come to the physician of his own volition, but must be brought by relatives or friends is rather suspicious; the true neurasthenic seeks medical advice without being urged, and is thoroughly conversant with his symptoms, even anxious to relate them. Lest he forget some of them, he keeps a diary in which his numerous troubles are faithfully recorded. The neurasthenic complains of loss of memory and of lack of mental concentration, neither of which is

complete or genuine, for he appears to remember everything he desires to remember and he can concentrate his attention on all matters of great interest to him; besides, errors of memory or attention are easily corrected. The paretic does not recognize errors in memory, and if he does, is unable to correct lapses by any amount of voluntary effort. The neurasthenic worries and is not satisfied with the physician's explanation of the case; the paretic's worries seem rather superficial and he is easily satisfied with any sort of explanation. The sudden onset of symptoms after some mental perturbation or shock favors the diagnosis of neurasthenia, while the insidious and slowly progressive development of symptoms speaks for paresis. Marked change of character, signs of loss or diminution in the ethical and moral spheres, marked hypochondriac depression without cause or explanation—these are symptoms favoring the diagnosis of paresis. The true neurasthenic, imagining himself insane, and uncertain in social intercourse, fears that which actually takes place in the paretic; but since he is really sane and knows how to behave in society, commits no offense against good manners and the rules of civilized society. Some of the symptoms foreign to neurasthenia but present in paresis are obstinate sleeplessness in spite of hypnotics, or the reverse, namely, an irresistible tendency to sleep anywhere and at any time. When these and similar symptoms appear in one who is supposedly a neurasthenic, the diagnosis of pre-paresis must receive serious consideration. Of course, a history of apoplectiform and epileptiform attacks, transient loss of speech, temporary monoplegia, or tongue paralysis, belongs to paresis and is against neurasthenia. The differential diagnosis between neurasthenia and general paresis was formerly made on clinical grounds only, and it therefore lacked the certainty of a laboratory examination. At present we are in a position to clear up grave doubts in diagnosis by the various tests: Wassermann, Nonne, Noguchi, Pandy and Lange. It is a rule admitting of no exceptions, that whenever a patient past middle age presents himself for the treatment of so-called neurasthenia coming on insidiously and without adequate cause, careful search should be made for the possible development of general paresis.

MELANCHOLIA.—The depressive form of paresis is distinguished from the mental depression of melancholia and the depressive phases of manic-depressive insanity by evidences of mental deterioration, defective judgment, failure of memory, lack of orientation as to time and persons, nonsensical delusions and entire absence of a true emotional background for the patient's words and acts. In melancholia, self-accusation and anxiety are rather sustained and accompanied by deep emotion, while in paresis these are transient in duration and devoid of feeling. The paretic has periods of exaltation interrupting his depression, while the melancholiac may have his good days, but never shows extravagant exaltation. In true depression there is motor retardation or inhibition, but no mental deterioration. A patient even in the stuporous states of manic-depressive insanity takes some notice of his surroundings; when threatened with a needle or pin he shows some concern and slowly

moves away the part threatened. The parietic is disoriented, takes little notice of his environment and does not react to painful stimuli. In addition, there are the characteristic physical signs, as Argyll Robertson pupils, disturbance of the deep reflexes, tremors, parietic speech, etc., all of which are absent in manic-depressive insanity. Finally, the Wassermann test on blood and spinal fluid is never positive in melancholia or manic-depressive insanity, while in general paresis there is almost 100 per cent. positive Wassermann.

MANIA.—The excited parietic may be mistaken for a case of mania or for one in the manic phases of manic-depressive insanity. In mania there is often a history of recurrent attacks, and delusions are within the bounds of possibility; in paresis the delusions are fantastic, improbable and ridiculous. The manic patient is more alert and quick to apprehend when his attention can be attracted, while the parietic is rather dull and depends entirely on suggestions from the surroundings. The maniacal patient is more or less consistent in his excitement, is not subject to sudden attacks of depression, while the excited parietic is more likely to have an accession of the most abject depression in the midst of a debauch of excitement. In the last analysis, the Wassermann test may offer valuable assistance.

CHRONIC ALCOHOLISM.—This condition occasionally produces a picture much like general paresis, and requires differentiation. The condition is a form of dementia and is often called pseudo-paresis. In this there may be found speech difficulties, pupillary irregularities, motor weakness, tremor, ataxic gait, exaggerated or absent tendon reflexes—in fact, many of the physical signs of general paresis and many of the mental ones. The speech disturbance in alcoholic dementia lacks the essentials of true parietic speech; there is no Argyll Robertson pupil; a history of chronic alcoholic indulgence is in evidence, and a syphilitic history cannot be obtained. In true paresis the symptoms are more or less persistent and the disease is progressive, while in alcoholic pseudo-paresis the symptoms change with the degree of elimination of alcohol. It must be admitted that the combination of alcoholism and general paresis is not infrequent and may lead to diagnostic difficulties, but the Wassermann and Lange tests will readily clear up the diagnosis.

THE PSYCHOSES OF INTERSTITIAL NEUROSYPHILIS.—These may take on the form of general paresis and present problems for differential diagnosis which gain in practical importance when it is recalled that the interstitial types of neurosyphilis offer better prospects for the therapist than the parenchymatous disease, general paresis.

Clinically we know that ocular palsies of the more or less permanent type belong to interstitial syphilis, while paresis of a transient type in the muscles of eye, upper or lower extremities are the distinguishing mark of general paresis. Optic neuritis with secondary optic atrophy belongs to interstitial neurosyphilis, while primary optic atrophy occurs in general paresis. In the last condition there may be found diffuse dulling of sensation, while in interstitial syphilis there may be localized sensory defects. Disturbances of speech and writing are never as ac-

ntuated as in paresis. Positive Wassermann with small quantities of spinal fluid, and the Lange paretic curve, may greatly assist in differentiating between the interstitial and the parenchymatous form of dementia.

SENILE DEMENTIA.—Senile dementia gives symptoms similar to those of general paresis and needs differentiation. The following criteria for senile dementia may be of use: The disease occurs at a rather advanced age, from 65 years on. Memory is lost for recent happenings, though it may be good for things long past. The patients are generally depressed and irritable, and believe themselves persecuted by their own relatives. Senile dementeds usually awaken very early in the morning and cause much commotion in their households, complaining of the ill-treatment accorded them by their kin. Though their delusions are mostly persecutory in character, occasionally they are of the grandiose type. Compared with general paresis, the course of senile dementia is slow; paretic speech is absent and Argyll Robertson pupil is relatively rare. The tremor in tongue, facial muscles, and in the hands, is present in paresis and absent in senile dementia. Lastly, the serological laboratory findings are positive in paresis and negative in senile dementia.

DEMENTIA PRÆCOX.—This condition is differentiated by the age differences; præcox occurring in younger individuals, while paresis, with the exception of the juvenile type, occurs mostly in persons past middle age. The physical signs of paresis are absent in the præcox group; there is good memory and orientation in præcox, while both are affected in paresis. Occasionally even *catatonic* symptoms are observed in general paresis, which of course leads to mistakes in diagnosis. In general paresis the catatonic symptoms are accompanied by greater insensibility and cloudiness and greater disturbance of memory. Both sets of patients may be extremely negativistic; and we then have to depend upon physical signs and laboratory tests for a diagnosis. Of the physical signs, the writer considers the Argyll Robertson pupil the most important one, for tremor, increased reflexes, epileptiform attacks, and dizziness may also occur in dementia præcox.

The *paranoid* forms of dementia præcox are distinguished by the patient's relative lucidity—he reasons better, though hallucinations are more frequent and delusions of influence are common in præcox and rare in paresis. Again, the physical signs and the laboratory findings will ultimately decide the diagnosis.

LEAD INTOXICATION.—Lead intoxication, especially that form called lead encephalopathy, may simulate the picture of general paresis: there are headaches, loss of memory and speech disturbances similar to those of paresis. There is, however, an absence of Argyll Robertson pupil, and usually there is proof of lead colic and lead palsy, and the dementia is moderate and non-progressive. Though Walter Timme has reported positive Wassermann in both blood and spinal fluid in a case of lead encephalopathy and cites Sir Thomas Oliver, Cyrus Field, and others, having published similar reports, this finding needs corroboration and the exclusion of syphilis. In lead cases the basophilic degeneration

in the blood of lead workers may greatly assist in diagnosis, also the gradual clearing up of the mentality after treatment of the lead condition by elimination. Besides, no one has as yet reported a positive Langley test in lead encephalopathy, and this test should definitely settle the diagnosis.

Treatment.—PROPHYLAXIS.—This is identical with the prophylaxis of syphilis in general and of neurosyphilis in particular. To the statements made under Interstitial Neurosyphilis and Tabes it is necessary to add the following considerations: The great preventive of general paresis next to thorough **antispecific treatment** is the **avoidance of excessive brain work, worry and responsibilities**. Emphasis must be placed on the importance of thorough antisiphilic treatment controlled by repeated Wassermann tests on blood and spinal fluid during the early stages of syphilis. According to Wile and Stokes, the fate of every syphilitic seems to be determined in the first months of infection; for it is then the nervous system receives its quota of spirochetes. The lesions during these early periods are transient, slight and readily amenable to treatment. It seems logical, therefore, to consider every patient showing involvement of the central nervous system in the early months as a case of potential tabes or paresis and entitled to the most effective treatment.

SPECIFIC THERAPY.—A positive diagnosis of general paresis was, until quite recently, the equivalent of a death warrant. It now means the most urgent call for the application of the most energetic antispecific treatment known. The methods of treating general paresis have undergone a slow evolution from pure empiricism to a more or less rational therapy of attacking the spirochetes in their hidden fields of activity. The non-specific method was evolved from the following clinical observations: Much improvement had been noted in the mental state of paretics after acute infections as pneumonia, erysipelas and febrile diseases generally; this was attributed to heightened leukocytosis accompanying those complications. The idea then suggested itself of inducing artificially this hyperleukocytosis by means of **sodium nucleinate injections** in ascending strengths. This method of treating general paresis is still on trial, having thus far yielded scant results.

Von Wagner's New Treatment of General Paresis.—This non-specific method has given excellent results in a number of cases and consists of the following: A non-treated case of tertian malaria is selected. While the malaria patient is in the midst of an attack of fever, 2 c.c. of blood are removed from his arm and injected into the skin of the paralytic patient's back. After a period of one to two weeks the injected patient develops chills and fever; the temperature may rise to 103–105° F. and even higher. The patient is permitted to pass through ten or twelve such attacks without being given quinin. Then quinin is administered in doses sufficient to check the malarial activity. And it is also at this time that the usual intravenous injections of salvarsan or neosalvarsan are administered.

Pilez reports that of 141 paretic patients treated in 1919–20, 51 have

completely recovered; 18 showed marked and persisting remission without being able, however, to return to their former occupations; 57 cases became stationary or showed an incomplete remission; 15 patients died. There is no parallelism in the degree and duration of the remissions on the one hand and in the serological reactions (spinal fluid) on the other. The best results are obtained in cases with maniacal excitement and in cases of the simple type of dementia paralytica. The hypochondriacal, presenile and catatonic forms of the disease show a less favorable response.

The remission, if it occurs at all, comes gradually, but finally becomes permanent. Not only general paresis, but other forms of neurosyphilis were thus treated. The results thus far obtained have awakened a new hope in the hitherto hopeless parietic.

In the correct evaluation of any form of therapy it is well to be reminded that general paresis has a natural tendency to remissions. According to Ball, 10 per cent. of all cases undergo remission of variable duration, and, 0.5-1 per cent. show complete and long continued remissions. It may thus happen that such remissions occur at about the time when treatment is begun and one is left uncertain as to what produced results—the remission or the treatment. It therefore behooves us to be rather conservative as to the value of any form of therapy. Nevertheless those who have treated a large number of cases must be accorded the right to an opinion as to the efficacy of certain methods of treatment. Because of the absence of results from the usual anti-syphilitic treatment by means of mercury and iodids, the medical profession had developed an apathy towards any kind of treatment of parietics. This fact retarded progress and led in part to the adoption of a wrong nomenclature for parenchymatous syphilis, namely, parasymphilis, metasymphilis and postsymphilis. Through the labors of numerous observers here and abroad, notably Noguchi, Nichols and Graves in this country, and Marie, Levaditi, Foerster and others in Europe, we know definitely that living spirochetes are present in the brain and also in the blood (Graves) of parietics. The inefficacy of **salvarsan** administered intravenously and of **mercury** by mouth, injection or inunction, is now easily understood, for the organism is not found near the blood stream, but in the gray matter at some distance from the blood-vessels. It has been shown by some that **arsphenamin** and certain other chemical substances administered intravenously scarcely—if at all—reach the ventricular fluid, whereas these same substances injected into the subarachnoid space readily reach the ventricles and through them are brought into communication with the perivascular and perineural spaces and clefts. Quite naturally the intraspinal injection route was seized upon as the most feasible method of reaching the brain parenchyma. Favorable results have been reported by most of those who have actually given this method a trial, though an occasional voice is heard against the futility and even harmfulness of the Swift-Ellis intraspinal method, and of the intracranial and intracistern routes. The results of intraspinal autoserosalvarsan injections have

been so encouraging for tabes that the same treatment is now applied in general 'paresis. Naturally, improvement has not been as marked in this disease as in cerebrospinal lues and tabes, which Swift explains by the fact that active inflammatory changes are often found in the region of the radicular nerves (in tabes), and the separateness of the dorsal roots may result in greater accessibility of the remedial agent. "It is conceivable," says Swift, "that if all the radicular nerves and dorsal ganglia were gathered in one area, as in the cerebral cortex, the arrest of tabes might be as unsatisfactory as the treatment of paresis."

Like Riggs, the writer has become impressed with the results obtained from intraspinal therapy, which are in striking contrast with our results from the older methods of administering salvarsan and mercury. Apart from all theoretical discussions as to why intraspinal medication may or may not be efficacious, the mere fact that patients who failed to respond to intravenously administered salvarsan have shown remarkable improvement under the administration of the Swift-Ellis method is sufficient recommendation for its employment in paresis. In hundreds of patients in whom intraspinal injections have been used, few or no untoward results have been reported, and this alone would indicate that the danger from its employment can only come from lack of skill and toxicity of the product. Besides, in a disease so utterly hopeless in its prognosis as general paresis, there need be no hesitancy in applying a remedy offering a promise of some measure of success.

Hammond and Sharpe have treated paretics by means of **salvarsanized serum** injected directly into the ventricles and they claim its superiority over the intraspinal and intradural method. In their first report they reach the following conclusions:

"1. In comparison with the deadly nature of paresis, the hazard of intradural treatment, by whatever method, is of little moment.

"2. On experimental and clinical grounds, both the subdural and intraventricular methods are superior to the intraspinal route in the treatment of paresis.

"3. From an experimental and theoretical standpoint, the intraventricular method is superior to the subdural route, and we believe it to be safer.

"4. The intraventricular method, with careful technic, and a due regard for the anatomy of the brain and the delicate nature of the tissues one is invading, is practically free from danger.

"5. If the freedom from unfavorable symptoms so far achieved in intraventricular injection can be maintained, it will be imperative to so treat paresis in its earliest stages, with greater chance for marked improvement and perhaps permanent arrest of symptoms."

In a second paper in which they report ten more cases treated by the intraventricular method, making thirteen in all, they state that the improvement noted in their cases has never before been obtained in

series of cases. There operative technic is simple and can be learned by a perusal of the original papers, mention of which will be found in bibliography at the end of this article.

F. Lautman describes a new method for intraspinal treatment of neural syphilis with mercury. His method consists "in pushing mercury to the point of tolerance, obtaining the blood-serum, reënforcing this by the addition of a solution of mercuric benzoate in normal saline, removal of as much spinal fluid as can possibly be obtained by lumbar puncture, and injecting intradurally the serum mercury preparation. The quantity of fluid injected should be less than the amount of spinal fluid withdrawn so as to create a negative pressure within the dural cavity." To saturate a patient with mercury, he gives daily inunctions of one-fourth of an ounce of fifty per cent. mercurial ointment, supplemented by tri-weekly intramuscular injections of any one of the mercurial salts. These procedures are continued until the first evidences of ptyalism appear (four breath, gingivitis, cramps, etc.), when 30 c.c. of blood are removed by venipuncture. The rest of the technic is similar to that used in the Swift-Ellis method:

"1. The blood is allowed to clot and the 10 or 15 c.c. of serum obtained is pipeted off and

"2. Centrifuged to remove all the blood-cells.

"3. One c.c. of a solution of one grain of mercury benzoate in 25 c.c. normal saline is placed in a clean test tube and boiled. If, on cooling, this solution becomes turbid, it should be discarded and another c.c. of the solution boiled up in the same tube. If this remains clear on cooling,

"4. The clear serum is added and the preparation is mixed well.

"5. It is then heated at 56° C. for half an hour and

"6. Administered, by gravity, at body temperature."

A method of treatment has been suggested and tried by Gilpin and Earley, which consists in **drainage of the cerebrospinal fluid** after the administration of mercury by inunction or salvarsan intravenously. They ask the question why arsenic and mercury, which produce good results in all other forms of syphilis, are practically innocuous in syphilis of the nerve tissue. The answer is given that after the administration of either drug by way of the mouth, skin, or blood-vessels, neither drug was to be found in the cerebrospinal fluid. Why? There are two answers: (1) The choroid plexus does not permit substances such as arsenic and mercury to pass. (2) Passage of the two substances into the cerebrospinal fluid takes place by osmosis, which is impossible when the pressure of the cerebrospinal fluid equals the pressure of the blood. Reasoning from this thought, they conceived the idea of reducing the pressure within the cerebrospinal cavity, in order to bring foreign substances through the capillary walls and is to secure the direct effect of arsenic and mercury on the nerve

tissue itself. Their technic is as follows: Patients are drained once weekly, and the quantity removed is as much cerebrospinal fluid as will flow from 20 c.c. to as high as 40 c.c. The patient is placed on his left side in bed. The sterilized needle is inserted at the second or third interspace. The first 5 c.c. of fluid are used for testing. The only bad effect of drainage is headache, which occurs in some patients. Some who have tried this method are quite enthusiastic over it, while others have not seen the benefits that are obtained from the Swift-Ellis method.

In the writer's opinion the intracerebral route, both intradurally and intraventricularly, should be left to the surgeon; the physician will do well to choose between spinal drainage and autoserosalvarsan intraspinal or intracistern injections. The writer prefers the combined mercury and Swift-Ellis method of treatment for early cases of general paresis.

The *tryparsamid* treatment described in the general section on Neurosyphilis under Treatment may after all become one of the recognized methods of treating general paresis. At the date of this revision, however, the author cannot conscientiously recommend its use. The danger to the optic nerve is such that it would overbalance any benefit that may otherwise be obtained.

Von Wagner's Malaria Treatment.—This method of treating general paresis, described on page 212 in a previous section of this article, is still in the experimental stage. At best it can only be applied in institutions for the insane and in hospitals. It is doubtful whether the individual physician in this country will ever be able to avail himself of this remedy even if it should prove valuable.

SYMPTOMATIC TREATMENT.—In addition to the treatment directed against the disease itself, symptoms as they appear in the individual case must receive attention.

For the attacks of excitement in paresis prolonged immersion in the **warm bath** at body temperature has been generally adopted by modern psychiatrists as an effective and humane substitute for physical restraint. Certainly the continuous warm bath is a civilized procedure compared with the *barbarous method of straps and strait-jackets*. In exceptional cases of extreme restlessness and excitement it may be necessary to administer small doses of **hyoscin hydrobromid** (grain 1/100) (0.00065 gram) hypodermically, or large doses of **sodium bromid** by mouth.

The treatment of attacks of melancholia and of hypochondriasis occurring in general paresis is the same as for depression from other causes, namely, the exhibition of **opium**, either the aqueous extract or the powder, in half-grain (0.0324 gram) doses, three times daily, and gradually increased to 1½ grains (0.1 gram) three times daily.

In the hypochondriacal cases, with the delusions principally directed toward the digestive organs, as when a patient thinks his tongue has

otted away and his stomach is made of glass, it may become necessary to resort to **tube feeding** or **nutrient enemata**.

For the better control and management of the epileptiform and apoplectiform attacks, **detention** in a private sanitarium or in a public institution is indicated, where the treatment will be the same as for epilepsy and apoplexy from other causes. It is an oft-repeated observation that the unexciting life of an institution is conducive to the prevention of the stormy episodes so common in the paretic's existence, especially early in the disease. Here also the outbreaks of violent attacks on the environment can be more readily subdued.

During the later and last stages of the disease, when the patient becomes more docile and manageable, he may be cared for at home by a competent nurse. It is well to instruct relatives that strict attention should be given to the most obvious rules of **general hygiene**. The paretic really must be treated as an invalid or child, for he loses all initiative. It will be necessary to keep the bed clothing perfectly dry and the skin should be cleansed with soap and water. The bony parts of the body must be padded so as to prevent bed-sores, which often terminate the patient's existence.

A constant danger in general paresis is the development of hypostatic pneumonia. We aim to prevent this complication by frequently **turning the patient from side to side**. It will also be necessary to guard against urinary retention and the development of cystitis. The *bladder* may be easily outlined by percussing the abdomen from below upward, and occasionally it may be emptied by **manual expression**, thus rendering catheterization unnecessary. In other instances regular **catheterization** must be resorted to, and immediately afterward the bladder is washed out with a weak solution of **boric acid** or **permanganate of potash**.

The *oral cavity* likewise requires the strictest hygienic care; mouth and teeth are cleaned after each meal, so as to prevent accumulation of additional poisons capable of aggravating the patient's condition. The *bowels* also come in for a share of attention, as paretics have been known to remain constipated for weeks without complaint.

And last but not least, it is the physician's duty once he recognizes the true condition, to warn the family against possible financial ruin. A paretic patient, being devoid of sound judgment, is easily recognized by money sharks and singled out as a victim. The result is that he enters into all kinds of get-rich-quick schemes with the inevitable consequences of losing the savings of a lifetime or his family's fortune.

To prevent such disaster, a conservator must be appointed to manage the patient's business and property. This can best be done by advising him to go away for a rest to a private sanitarium. Of course, relatives are not always convinced that such a course of action is necessary until too late to save even a fragment of the patient's property, for it is hard to believe that under an apparently normal exterior an individual is entirely devoid of judgment. However, it is our duty to

present the facts in the most forceful manner and to obtain the family's consent to relieve the patient of all social and financial responsibilities.

Prognosis.—Ever since the disease was first recognized the prognosis has been considered decidedly unfavorable. Death occurs in a majority of cases within three years from the onset of the disease. Only a small proportion of patients live five or six years after the full development of symptoms. According to Charles L. Dana, there are "cured" or arrested cases of paresis, of which he has reported a number. It was claimed by others that his cases were not true paresis but merely resembled that disease clinically—were so-called pseudo-paresis. To this Dana has replied more than once and each time he is more firmly convinced that his view is correct. He supports his contention by further reports of cases previously described as cured. It appears to the writer that no one has disproven the diagnosis of any of his cases. In the future we shall have *positive* proof as to our cures only when all the newer differential tests, including the Lange and Luetin reactions, shall be systematically applied in every case before beginning treatment. In the writer's opinion it is a mistake to pronounce a patient doomed when the diagnosis of general paresis has been made, and while convinced that lengthy remissions are possible even with insufficient treatment, it appears quite probable that we shall be able to change the prognosis to a more favorable one by the application of energetic treatment. In a number of cases coming under the writer's personal observation he has seen remarkable improvement and arrest of symptoms to a degree considered "cures" by relatives and friends. Though the number of patients who were thus benefited by treatment is not large, reports are constantly reaching us from various sources establishing the fact that undoubted cases of paresis are making splendid progress under the intraspinal and intracranial methods of treatment. Of course, no one is deluded as to the outlook of advanced cases of tabes or paresis, which scarcely show lengthy remissions under the best treatment. In such cases nothing more is required than custodial care; all efforts toward a cure are misdirected and a sign of poor judgment. However, in the early cases, when a supposed neurasthenic in the pre-paretic stage of general paresis comes to us, at a time when the diagnosis is made with some difficulty, treatment energetically pursued gives the best results. The prognosis, therefore, depends largely upon the stage of the disease. In addition, the patient's tendencies and his general make-up are to be considered. Those who have dissipated all their lives and who are derived from neurotic stock seem to grow worse under any form of therapy. It is the writer's opinion that, given a patient in the earlier stages of the disease and one who has lived a fairly good life as regards excesses in *baccho et venere*, and who is in a condition to avail himself of the modern treatment by intraspinal, intracranial, or spinal drainage methods of treatment, the prognosis is by no means hopeless. On the contrary, upon a perusal of the recent literature one gains the impression that the parietic's outlook for improvement and cure has become better than ever before.

Pathology.—In recent years the pathological anatomy of general paresis has been worked out to a degree that may be considered pathognomonic for that disease. Beginning with the skull, it may be noted that occasionally hyperostoses and exostoses with thickening of the tables have been observed. The important pathological changes are found in the membranes and the cerebral tissues.

The dura is more or less adherent to the skull and strips with difficulty from the brain substance. There are evidences of pachymeningitis interna and hematoma of the dura mater in the form of freshly organized material and laminated layers of recent and old blood-clots. In addition, numerous subpial hemorrhages may be observed and the leptomeninx is thickened; milky deposits can be easily made out along the blood-vessels.

Microscopically there are found proliferated connective tissue and numerous lymphocytes and plasma-cells, also mast-cells and so-called gitter-cells. The convolutions are atrophied, especially in the frontal lobes, less so over the parietal lobes. The pathological changes are less marked at the base and barely noticeable over the occipital lobes. The veins are much dilated, especially in the acute forms of general paresis.

The connective tissue of the cortex, the neuroglia, shows important and extensive pathological changes which Weigert has described as characteristic for destruction of nerve parenchyma. As in other tissues, in which connective tissue replaces destroyed parenchyma, neuroglia takes the place of the proper parenchyma in the brain. One of the principal characteristic pathological changes in paretic brains is the abundance of neuroglia tissue—glia cells and glia fibers—an expression of much parenchymatous brain destruction.

In addition to the changes described, the blood-vessels are affected; there is proliferation of the interstitial cells with division of nuclei. This is best seen after apoplectiform attacks when new vessels form and the cells proliferate into the surrounding tissue, forming new interstitial cells and fresh capillaries. On sectioning the brain one sees numerous capillary vessels, whose walls have undergone hyaline degeneration; likewise dilated vessels and capillary hemorrhages. The characteristic change in the vessels consists in a large accumulation of cells in the dilated adventitial lymph sheaths, to an extent observed only in this disease. The cells are not ordinary leukocytes, but lymphocytes and plasma-cells, as described by Nissl and Alzheimer. Plasma-cells are circular or ovoid cells, whose plasma stains irregularly and which show a bright area toward the center. The nucleus is not exactly in the center of the cell and its border is usually surrounded by a number of thick chromatin bodies and one or more nuclei. The plasma-cells are supposed to have originated from adventitial connective tissue, according to some, while others consider them as metamorphosed blood-cells. The plasma-cell infiltration must stand in some relation to the paralytic changes, for in general paresis they are plen-

tiful, while such cells are not encountered experimentally and have not been observed in the brains of normal individuals.

Mast-cells, often seen in cases of meningo-encephalitis, are never absent in paresis. Their place of predilection in general paresis is in the frontal lobe, becoming less frequent as we go posteriorly, and being altogether absent in the occipital lobe.

Another type of cell observed in general paresis is the so-called rod-cell (Stäbchenzelle) described by Nissl. This is a long, rod-shaped cell with a small body, a faintly stained nucleus and several nucleoli, which seem to extend beyond either end of the cell-body. This variety of cell is most frequently found in the vessels and runs parallel to the long axis of the large nerve-cells, but these cells are found more irregularly placed and occupy deeper layers. Nissl and Alzheimer think they are derived from blood-vessels, while Sträussler and Cerletti consider them of glial origin. Isolated rod-shaped cells have been seen in other diseases of the cortex, but never in such profusion as in general paresis.

In addition to the finer microscopic changes in the cortex there are occasionally seen small areas of softening. It has been stated by Kraepelin that gross focal lesions are but rarely encountered, but Macfie Campbell in his thesis has shown by a description of his material that focal lesions are not so rare.

The basal ganglia, central gray matter, and cerebellum, also present degeneration of the nerve-cells and fiber tracts. Similar changes to those seen in the cortical cells have been observed in the cranial nerve nuclei of the medulla.

In the spinal cord the tracts most frequently involved in almost all cases are the posterior and lateral columns. Even the peripheral nerves occasionally show degeneration.

Is it possible to diagnose general paresis with any degree of certainty from the pathological anatomy alone? For a long time this question had to be answered in the negative. We may now state that the diagnosis has become more nearly certain. It is true that each single change described may occur in other forms of brain disease, but the *ensemble* of the pathology as outlined for various tissues and portions of the brain permits a probable diagnosis of general paresis.

Referring to the relative importance of the individual pathological alterations, it appears that an accumulation of plasma-cells in the arterial walls is most important for a diagnosis of paresis. Next in importance rank the numerous rod-shaped cells, endothelial proliferation and new capillary formations, the extensive destruction of nerve-cells proper and the proliferation of glia tissue in the deeper cortical layers. Besides, the peculiar depression and the diffuse changes in the pia and cortex, the favorite location over the frontal lobes and a relative freedom from disease of the occipital lobes are points of importance in the pathological differential diagnosis between paresis and syphilitic meningo-encephalitis or tuberculous meningitis. A point worth mentioning is that the brain changes occurring in the latter conditions are the result of concomitant meningeal alteration, and not primary, as in general

paresis. Finally, the presence of atrophy and shrinking of the brain cortex also aids in the diagnosis of general paresis. This disease destroys more of the cortex and leads to greater vascular proliferation than any other, such as dementia præcox, senile dementia, or idiocy. In senile dementia, for instance, numerous cells and fibers undergo destruction, but the general structure of the cortex of the brain remains intact. Perhaps the only other disease which creates similar pictures is cerebral arteriosclerosis.

What is called the *pathology* of general paresis is only a small part of the subject, namely, pathologic anatomy. The real pathology is still to be written. Why do the spirochetes affect the parenchyma of the brain in some cases and not in others? According to most authors, the spirochetes take their final position during the period of secondary septicemia and the future course of the disease is determined by their early localization. The spirochetes have been demonstrated in the brain cortex, the spinal cord and the spinal fluid. It appears probable that certain strains of spirochetes possess localizing powers for certain situations. Nichols, from his experiments and from observation of cases, formulates the theory of the inhibitive effect of an active lesion. In the human this experiment is made by nature thus: Given a case of secondary syphilis with invasion of the nervous system, as long as the skin lesions are active, symptoms rarely occur in the nervous system. No sooner have the skin lesions healed, when, if some spirochetes have been left in the nervous system, the inhibitory effect of the skin lesion is lost and a lesion flares up in the nervous system. The same theory might explain why systemic lesions are rarely seen in general paresis. Their continuous activity here has an inhibitory effect upon the few remaining foci in other organs. If this pathology and the explanation are correct, a practical inference would be that the most energetic and relentless warfare against the spirochetes must be conducted so as to exterminate the entire brood in every organ.

(c) SYPHILITIC PROGRESSIVE MUSCULAR ATROPHY

Introduction.—Muscular atrophy is not uncommon in neurosyphilis. The cause may be either spinal root involvement from meningeal exudates, or else anterior horn compression from gumma. In some instances arterial thrombosis may produce shrinking of the anterior horn cells with consequent muscular atrophy. These varieties of atrophy belong to interstitial syphilis. There are cases of muscular wasting in connection with tabes and late cerebrospinal syphilis which may be considered complications of these disorders. The type with which we are now concerned is that form of parenchymatous neurosyphilis in which progressive muscular atrophy appears as an independent syphilitic affection.

Etiology.—Since the etiology is the same for all forms of syphilis, namely, the entrance of the spirochetes into the tissues involved, it is only necessary to state that spirochetes have actually been found in

the anterior horns of the spinal cord. The question, why the anterior horn cells should suffer in certain individuals and not in others, may perhaps be answered by the unsatisfying reply of predisposition and stress.

Symptomatology.—The disease usually has a subacute beginning, requiring one or two months for its development and reaching its height perhaps in three or four months. In some cases the disease comes to a standstill before the end of a year and the symptoms either remain quiescent or improve during one or two years. After this time there is usually a recrudescence of symptoms which indicates involvement of neighboring groups of cells. There may be no further extension of the disease processes after the second attack, but sometimes the disease continues to progress in an upward direction, implicating the bulbar nuclei and terminating fatally. The symptoms of paralysis or weakness precede the flaccid atrophy, but otherwise the disease follows the course of non-specific muscular atrophy. The upper extremities are affected in the shoulder-girdle, and eventually the forearms and hands show extensive atrophy, or the wasting may begin in the small muscles of the hand and involve the higher parts later.

The following case from the writer's hospital practice embodies most of the symptoms ordinarily found in this disease.

CASE XXIII.—The patient is an American, 40 years of age and has a good family and personal history. He admits having had a chancre 20 years ago, which was insufficiently treated, not having been recognized as syphilitic. He entered the hospital because he is practically helpless in his upper extremities, his arms and hands dangling about the body as lifeless members.

Examination.—The objective examination reveals a neck without proper support; the head and neck can be passively flexed and extended without the patient being able to offer the slightest resistance. There is marked wasting of the anterior neck muscles, but the posterior group seems to be fairly well preserved. Beneath the clavicle on either side there appears a hollow space where the pectoralis major should be, the pectoralis minor having been left intact. The absence of these muscles can be easily demonstrated by the patient's inability to adduct his arms and forearms by pressing his palms one against the other; the examiner can force them apart with a minimum of resistance on the part of the patient. The deltoid muscles are practically lost on both sides, so that the patient is unable to raise his arms in a forward, lateral or posterior direction. The supraspinati and infraspinati muscles on both sides have disappeared, but the triceps muscles are practically preserved. The biceps is paralyzed and atrophic on both sides, more markedly on the left. The entire group of forearm muscles has flattened out and is parietic, especially the extensors, giving a typical bilateral wrist-drop. The thenar eminences have disappeared, while the hypothenars are but little affected. Both median and ulnar territories are unequally affected, the ulnar side being somewhat spared. There is

practically no power in the right hand and very little in the left. Thus the muscles of the abdomen are uninvolved, likewise those of the spine below the neck, also the pelvic girdle and the lower extremities.

Reflexes.—Wrist and elbow jerks are absent. The triceps is present, but reduced. Knee and Achilles jerks are normal. An examination of the eyes reveals what is perhaps of greater importance for a diagnosis than a positive Wassermann, namely, an insignificant light response in the left eye and practically none in the right eye.

Summary.—The disease picture is one of progressive muscular atrophy of the spinal type due to involvement of the motor cells in the anterior horns, there being no sensory disturbances whatever. The entire condition developed during a period of four months.

Differentially in this case we must distinguish between the ordinary type of poliomyelitis and the syphilitic variety, because there are differences in prognosis and treatment. The patient having passed through a syphilitic infection twenty-two years ago, the case probably now belongs to parenchymatous neurosyphilis, not the interstitial type. There is degeneration of the anterior horn cells in the cervical cord with consequent shrinking and atrophy, which has resulted in a flaccid paralysis of the muscles supplied by that part of the cord. The disease belongs to the variety named syphilitic chronic poliomyelitis or syphilitic progressive muscular atrophy. There is no doubt as to the diagnosis—the history of chancre 20 years ago; an Argyll Robertson pupil and positive Wassermann on the spinal fluid are sufficient for a definite diagnosis.

LABORATORY FINDINGS.—These are nearly the same as for tabes—lymphocytosis, positive Wassermann on spinal fluid, an increase of globulin by Nonne and Noguchi tests. In a number of cases, as in tabes, the Wassermann tests will be negative.

Diagnosis.—This will be largely made from the history of specific infection, the finding of the well-known clinical signs of neurosyphilis in various parts of the body and of positive laboratory tests. The disease must be differentiated from progressive muscular atrophy of the non-specific type. Clinically there are no points of difference, except perhaps the frequent presence of pain and paresthesia preceding the onset of the muscular weakness, which constitutes a symptom favoring the syphilitic variety. From the interstitial type of muscular atrophy this form can be differentiated by the time element, parenchymatous neurosyphilis appearing from 10 to 20 years after infection, while the interstitial form appears within 2 to 5 years after chancre.

Treatment.—The treatment of syphilitic progressive muscular atrophy is not essentially different from that of tabes. The **Swift-Ellis method of salvarsan therapy** seems to be especially indicated for this disease, besides the **intensive mercurial treatment**. In addition, the faithful systematic application of **mild massage and galvanism** to the affected muscles should do good; violent massage, however, must be avoided, as it may do harm. Of course, the **hygienic rules** laid down for all forms of neurosyphilis are here indicated with increased force.

Prognosis.—The prognosis as to life is not bad. Patients may live many years with this disease, even if untreated, unless the process extends to the medulla. Improvement from treatment has been noted in so many cases that we are led to believe that many of the cells have

been saved from complete destruction by appropriate treatment. Of course the outlook is better than in general paresis and, if it were not for the muscular disabilities, is perhaps a shade better than in tabes.

Pathology.—The pathology as described by Spiller, Dana, and Mott is practically the same as in ordinary progressive muscular atrophy. The anterior horns show considerable shrinking and the cells have disappeared in numerous places. The vessels have been found thickened and the connective tissue increased. A moderate amount of degeneration has also been noted in the anterior roots of the several portions of the cord, depending upon the part of the cord affected. In addition, some round-cell infiltration into the pia has been described, as well as small hemorrhages within the gray matter.

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CHAPTER XIII

TUBERCULOUS MENINGITIS

By JOSEPHINE B. NEAL, M.D.

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Definition.—The name of the disease sufficiently defines it. It is an inflammation of the pia mater due to the tubercle bacillus.

Etiology.—**PREDISPOSING CAUSES.**—More cases are said to occur in the *late winter* and *early spring* months than at any other season. Holt, in analyzing 218 cases in New York, found the maximum number in April; Meyers, in January, according to an analysis of 105 cases in Boston. The author's own records for 432 cases are as follows:

January	—	36	July	—	49
February	—	40	August	—	29
March	—	46	September	—	30
April	—	42	October	—	18
May	—	51	November	—	15
June	—	49	December	—	27

This does not bear out the common opinion, as the maximum numbers for these cases fall in May, June and July. This fact is doubtless due to respiratory diseases, so prevalent in these months, developing latent tuberculous processes in the lungs or bronchial nodes. Sometimes these foci are *developed by other diseases*, grip, measles or whooping cough. Certainly it seems to the author that many of the cases within her experience give a history of an attack of measles within a year. Meyers says a past history of measles was given in 26 per cent. and whooping cough in 22 per cent. of 105 cases. Not only may these diseases break down an old tuberculous lesion, but they also lower the resistance of the individual. Dopter and others report cases of tuberculous meningitis developing secondary to other forms of meningitis, particularly epidemic (meningococcic). They attribute it to the lowering of the local resistance of the meninges by the primary infection. Sex has no influence on the development of the disease.

The *age* is of great importance. The disease is comparatively rare under the age of six months. Holt gives three cases out of 410 under three months. The author has found four cases out of 547 under three months. The greatest number of cases occurs between the ages of six months and two years. Holt gives the following data:

Under 1 yr.	162
1 to 2 yrs.	149
2 " 5 yrs.	76
5 " 9 yrs.	17
9 " 16 yrs.	6

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The author's distribution according to age has been as follows:

Under 3 months	4
3 mos. to 6 mos.	17
6 mos. to 1 yr.	63
1 yr. to 2 yrs.	154
2 yrs. " 3 yrs.	72
3 yrs. " 5 yrs.	77
5 yrs. " 10 yrs.	84
10 yrs. " 20 yrs.	42
Over 20 years	34

547

The question of *trauma* sometimes arises. It does not seem to be of importance in view of the fact that all children, more or less, receive bumps on the head without subsequent ill effects. Of course, trauma of a joint, for instance, may develop an old tuberculous focus, thus disseminating the bacilli and thereby being instrumental in causing a meningitis.

In regard to *hereditary tendencies*, *social conditions* and so on, the author quotes from Hutinel and Voisin:

"We find first a predisposing cause which exceeds all others in importance: the neuropathic heredity. This must be considered in its largest sense. The parents whose children die of tuberculosis are not always the insane, the epileptics, the hysterical, or the alcoholics; they are often highly intelligent individuals, where cerebral activity has been maintained at an excessively high tension and who have acquired from this mental over-stimulation an exaggerated impressionability or a certain degree of neurasthenia. As for the children, some present stigmata of degeneration; others have had convulsions in their early years, but most are very active children, of a precocious intelligence, whose large skulls and highly developed cerebral activity contrasts with a physical frailty; sometimes they are very beautiful children, in whom it is difficult to suspect a tuberculous taint.

"They belong more often to the well-to-do classes than to the poor, and they are much more numerous in large cities than in the country."

The author's own experience has been largely among the poor of New York City and this quotation is given because the experience of the writers has included a wide social range.

EXCITING CAUSE.—The exciting cause is, of course, the tubercle bacillus. Its morphology and distribution are discussed elsewhere and the author will not repeat them, except as they bear on the subject in hand.

Tuberculous meningitis is practically always *secondary to some tuberculous lesion elsewhere*, though it is often impossible to locate it clinically. A careful autopsy reveals such a focus in nearly every case. The mediastinal or abdominal lymph-nodes seem to be the most common site to be located. Parrot, Hutinel and Kuss, however, state that these lymph-nodes are manifestations of a tuberculosis secondary to an acquired infection, of which the lung is the portal of entry in nine cases out of ten, though it may also enter by way of the intestinal tract, the pharynx or the skin. As Hutinel and Voisin point out, however, "The division of tuberculous meningitis into primary and secondary is of only relative importance. A meningitis that has all the signs of a primary disease may be considered clinically as primary when the local tuberculosis from which it proceeds is sufficiently benign to remain latent, but once established it differs little from that which develops in the course of a tuberculous infection sufficiently well-marked to be easily recognized." While the infection of the meninges in certain cases may take place by way of the lymphatics or by direct extension, it is believed, in the majority of cases, to take place through the bloodstream.

As regards the *method of acquiring the infection*, a history of direct exposure is often unobtainable, in the author's experience. In Meyer's cases, there was no known exposure in 73 per cent. of the histories. Of course, it is only too easy for a child to be exposed to tuberculosis by unrecognized or unadmitted cases in relatives, friends or neighbors. Milk from tuberculous cows is another possible source of infection, but this seems to be operative in only a small percentage of cases. Holt reports 32 cases worked out by Park and Krumwiede, of which 30 showed bacilli of the human type, one of the bovine type and one of both types. Park and Williams give the following figures from their own and reported cases: tuberculous meningitis, secondary to tuberculosis of alimentary origin, 1 case human, no bovine, in children of from 5 to 16 years; 5 cases human, 10 bovine, in children under 5 years; in tuberculous meningitis, other than the preceding, 13 cases human, no cases bovine, in children of from 5 to 16 years; 104 cases human, 6 bovine in children under 5 years. In a series of 48 cases recently worked out by Mr. Novick in connection with the meningitis work at the Research Laboratory, 45 were of the human type, and three of the bovine type.

Including the author's figures with those of Park and Krumwiede, we have a total of 187 cases, of which 19 were of bovine type and 168 of human type.

Symptomatology.—**CLINICAL HISTORY.**—*Period of Incubation.*—The exact incubation period is not known. And, of course, the question would arise as to whether the period of incubation should be counted from

the time when the primary infection is received or from the time when dissemination from this primary focus takes place. Most writers agree that the period of incubation is probably long—several weeks or months. Still cites two cases in which the time of exposure and probable infection were known and preceded the onset of symptoms in one instance by a little over two months, in the other by a few weeks.

Mode of Onset.—The period preceding the onset of tuberculous meningitis is, in the great majority of cases, free from symptoms of a tuberculous infection, and the patient is in apparently the best of health when the disease begins to show itself. In a few cases only do the meningeal symptoms develop in the course of a pneumonia or other acute illness. In these instances, the onset is abrupt. The author recalls one case in which a child suffering from pneumonia began so suddenly to develop meningeal symptoms, and in which the symptoms were so well-marked, that the author felt sure the case was purulent meningitis secondary to the pneumonia. In another instance, a woman was sent to Willard Parker Hospital with a diagnosis of influenzal meningitis, the meningeal symptoms having developed rather suddenly following an attack of influenza. In some other cases, the onset is sudden, but usually it is gradual and is characterized by drowsiness and disinclination to play, fretfulness and irritability.

Headache is an early symptom in those old enough to be questioned. Hutinel and Voisin, and Castaigne and Paiseau make quite a point of a prodromal period, with nutritional disturbance, loss of weight, disturbed sleep, etc. Such a history has been the exception, not the rule, in the author's cases. Indeed we have been greatly surprised to see how well-nourished and healthy most of the children have seemed and we are never called in until what would be described as a prodromal period is well passed.

The early symptoms, then, are about as follows: Vomiting and constipation are found and headache in children old enough to indicate it. The vomiting is usually not severe and may be associated with the taking of food, though it is commonly projectile. The headache, especially in older children, is sometimes excruciating, so that they cry out every few minutes with the pain, and give a picture of very acute suffering, quite different from the somewhat somnolent condition that we commonly associate with tuberculous meningitis. There is drowsiness and indifference, following fretfulness and irritability. There may be convulsions. The hydrocephalic cry and grinding of the teeth are other evidences of irritation of the central nervous system. One of the earliest and most constant signs is an irregularity of the pulse. It is a curious irregularity more in force than in rate, a weakening of an occasional beat, and is entirely different from the sinus arrhythmia so common in children. The pulse is usually fairly rapid. In only a few instances is there a very slow pulse, below 60. The irregularity becomes more pronounced as the disease progresses. The respirations are usually irregular, when the condition is well-established and in most cases Cheyne-Stokes breathing is present at some time. The temperature is usually low, under 101° F. (38.3° C.), irregular, at times sub-

normal. At the end, it often rises to 105° or 106° F. (40.5° or 41.1° C.). It is evident that the course will be modified if a bronchopneumonia, for instance, is also present. In some cases the temperature is elevated throughout the course, without clinical evidence of a bronchopneumonia.

PHYSICAL FINDINGS.—The increased pressure of the cerebrospinal fluid is evidenced by a bulging fontanel or by the presence of Macewen's sign after the fontanel is closed. This sign is elicited by percussion over the lateral ventricle. A tympanitic sound and a feeling of elasticity to the percussing finger indicate tension due to the increase in cerebrospinal fluid. The sign is quite reliable to an experienced observer. No sign is infallible. The author has, on several occasions, found an increase in the spinal fluid where the fontanel did not bulge.

There is rigidity of the neck in most cases over 2 years of age. In children under two years, rigidity is often difficult to determine on account of the active resistance of the child, which leaves one in doubt. The rigidity is only moderate, in contrast to the marked retraction that is often present in purulent meningitis. The Kernig also is much less marked than in purulent meningitis. Indeed, Meyers reports that it was absent in 71 per cent. of his cases. The author's own experience does not confirm its absence in so large a percentage of cases, but it is certainly slightly more moderate in a much greater number of cases than in the purulent form, and in very young children it is usually somewhat indefinite.

The Brudzinski neck sign is frequently present. It is usually considered as of more value than a Kernig or rigidity of the neck in young children. Meyer found that it is usually absent in patients over 5 years of age.

The Babinski sign has never seemed to the author of great importance. It is so irregular in children under 2 or 3 years of age that its presence or absence means little in these cases. As it indicates a lesion of the upper motor neuron, its presence in older cases would simply mean that the cerebral cortex was involved in the inflammation as well as the meninges.

The reflexes usually show changes. The pupils, in rare instances, may be equal and react normally to light within a few days of the end. Usually, however, when the disease is well advanced, the pupils are dilated, often unequally, and respond sluggishly or not at all to light. The pupils are more frequently unequal in tuberculous than in other forms of meningitis. The knee jerks may be equal and active or even exaggerated, early in the disease; later they are most frequently diminished or absent, but they are usually equal on the two sides.

Paralysis, especially of the muscles of the eye or face, is common. Meyer found some form in 43 per cent. of his cases. A paralysis of the arm or leg or a hemiplegia may occur, however, and lead one to suspect a poliomyelitis. The paralyzes are often transitory, or show frequent variations in degree. Convulsions may be an early symptom, especially in the younger children, and they sometimes occur at the end.

TABLE 1.—SYMPTOMS OF TUBERCULOUS MENINGITIS (425 CASES)

Symptoms	Under 2 Years	2-10 Years	10-20 Years	Over 20 Years
Onset				
Sudden.....	43	59	9	4
Slow.....	122	139	21	10
Indefinite.....	6	9	...	3
History of T. B. in family.....	13	7	1	1
Indefinite.....	154	193	25	...
Definitely negative.....	4	7	5	1
Evidence of other T. B. foci....	6	9	6	8
Headache				
+.....	?	126	30	14
-.....	?	48
Indefinite.....	...	33	1	3
Vomiting				
+.....	138	165	27	5
-.....	24	30	3	11
Indefinite.....	9	12	1	2
Convulsions				
+.....	92	113	8	1
-.....	64	77	21	13
Indefinite.....	8	17	2	5
Cerebration				
Stupor.....	149	179	19	8
Delirium.....	3	12	4	3
Normal.....	2	6
Irritable.....	11	15	4	4
Indefinite.....	6	5	4	2
Fever				
Below 100° F.....	36	58	3	4
100°-103° F.....	107	139	21	8
103°-105° F.....	9	4	5	2
Indefinite.....	19	7	2	3
Pulse				
Regular.....	43	44	10	8
Irregular.....	67	80	7	4
Indefinite.....	61	81	14	5
Rapid.....	83	69	11	3
Slow.....	5	24	9	8
Moderate.....	36	43
Indefinite.....	47	71	11	6
Respiration				
Regular.....	21	42	7	9
Irregular.....	64	60	6	1
Cheyne-Stokes.....	30	29	1	...
Indefinite.....	46	76	17	7

TABLE 1.—SYMPTOMS OF TUBERCULOUS MENINGITIS (Continued)

Symptoms	Under 2 Years	2-10 Years	10-20 Years	Over 20 Years
Reflexes				
Increased.....	33	25	3	3
Normal.....	37	55	7	6
Diminished or absent.....	45	86	15	4
No information.....	45	41	6	4
Unequal.....	11
Paralysis				
Strabismus.....	26	39	3	2
Other.....	26	17
Macrowen.....	141	188	17	9
Duration				
Under 2 weeks.....	31	10	3	1
2-3 weeks.....	56	69	11	4
3-4 weeks.....	47	88	1	1
4-5 weeks.....	9	17	3	1
5-6 weeks.....	1	5	...	1
Over 6 weeks.....	3	10	4	2
Indefinite.....	24	8	8	7
Pupils				
Equal.....	58	87	16	11
Unequal.....	25	24	1	2
Indefinite.....	88	96	14	4
Reaction to light.....	43	69	15	10
No reaction to light.....	66	72	8	2
Indefinite.....	62	56	8	5
Rigidity of neck				
Slight.....	86	83	10	6
Moderate.....	45	77	15	9
Marked.....	7	14	1	...
Absent.....	12	8
Indefinite.....	21	25	5	2
Kernig				
Slight.....	66	57	9	3
Moderate.....	36	83	13	7
Marked.....	...	19	3	0
Absent.....	34	24	1	2
Indefinite.....	34	24	5	5

The tache cérébrale is of little value since it is often present in normal people, but it has been mentioned in literature so often that it has become somewhat classic. There may be delirium at some stage, usually of the low, muttering type.

As the disease advances, the drowsiness and apathy deepen into coma from which it is impossible to rouse the patient. Swallowing becomes difficult or impossible. A striking feature of the progress of tuberculous meningitis is the fact that remissions occasionally occur, even lasting for a few days, and one is led to hope that a mistake has been made in the diagnosis. Lumbar puncture is followed by improvement lasting for a few hours, unless it is very near the end.

The accompanying table (Table 1) shows the symptoms observed at the time the author's cases were examined, and the history of the cases up to that time. Of course, such a chart, prepared from single observations for the most part, gives, as it were, a cross-section diagnosis of the disease rather than a complete clinical picture. However, we are expected to make at least a tentative diagnosis of a case at our first examination, and for this reason the chart may be of value. The author regrets that "no information" appears in so large a number of cases. Two extenuating circumstances should be considered, in addition to the difficulty inherent in human nature of keeping adequate records; in many of the author's cases it was difficult to obtain really reliable histories, and at times the pressure of work has prevented the keeping of records.

LABORATORY FINDINGS.—While tuberculous infections are, as a rule, unaccompanied by leukocytosis, it may be associated with tuberculous meningitis. According to Holt, the blood-count depends on the stage of the disease. At first, the leukocytes may be normal or even diminished, while the last week there is a leukocyte count averaging 29,600, with a polymorphonucleosis of from 70 to 85 per cent.

Of course, tubercle bacilli may be found in other lesions. Holt reports that in his experience tubercle bacilli have been found in the sputum in nearly half of the cases, although in most of them there was no well-marked evidence of pulmonary involvement.

The most valuable aid that the laboratory can give in the diagnosis of tuberculous meningitis is in the examination of the spinal fluid. Indeed this will usually determine the diagnosis at once. If the first fluid is withdrawn at an early stage of the disease and the findings are inconclusive, further examination may be necessary.

The spinal fluid in tuberculous meningitis is increased in amount and is usually clear, though, rarely, it may be hazy, in cases of an unusually acute course. When a tube of the fluid is allowed to stand, a fibrin web forms, but this may occur in other conditions and is not pathognomonic of tuberculous meningitis.

The cells are increased, counts are given ranging from 25 to 1,000. We find it sufficient to centrifuge the fluid and examine the sediment, after it is fixed and stained with Ziehl's stain. We then mark the cells as slightly, moderately, greatly or very greatly increased, and note the relative proportion of mononuclears and polymorphonuclears. The actual cell-count has no diagnostic value; the counts of poliomyelitis epidemic encephalitis and tuberculous meningitis overlap, and these are the chief conditions from which the diagnosis must be made. The cells usually show a preponderance of mononuclears, from 80 to 100 per cent. In rare cases, especially in the hazy fluids, there is a preponderance of polymorphonuclears. Fortunately, in these cases, the tubercle bacilli are usually easy to demonstrate, or the question of some purulent form of meningitis, as well as of poliomyelitis, would arise.

Two methods of demonstrating the tubercle bacilli are in use—teasing out the web, and obtaining a sediment by prolonged centrifuging (an hour at high speed). As we usually begin the examination of the

fluid at once, it is more convenient for us to centrifuge and examine the sediment. Hemenway found the bacilli in nearly 100 per cent. of cases using the web, and she notes that the last portion of the spinal fluid is the best to examine for organisms. Whether or not the organism is found depends really on the time and patience at the disposal of the examiner.

If tubercle bacilli are not satisfactorily demonstrated, 5-10 c.c. of the fluid should be injected subcutaneously into the groin of a guinea pig. At the end of a month, the pig should be injected in the axilla with 1 c.c. of crude tuberculin diluted to 3 c.c. with normal saline. Usually, if tuberculous, the pig is dead the next morning. At autopsy caseous glands or tuberculosis in the spleen, or both, are found. Killing the pig by tuberculin makes the diagnosis of tuberculous lesions practically certain, even if the lesions themselves are rather indefinite.

The albumin and globulin are increased, sometimes moderately, sometimes very greatly. The presence or absence of glucose as shown by the reduction of Fehling's is a very important aid in the diagnosis. Very early in the disease the reduction may seem practically normal, but as the disease advances, the reduction almost invariably becomes distinctly less or even absent. This is in contradistinction to the fluids of poliomyelitis or encephalitis or syphilitic involvement of the central nervous system, where the reduction of Fehling's will remain normal. If there is a question of syphilis, of course, a Wassermann will help to clear up the diagnosis. The gold chloride curve is also of value in differentiating these two conditions.

Some time ago, we were able to have some gold chloride curves made from various types of fluids. The curve of 13 fluids from tuberculous meningitis was 1112221000.

Seventeen fluids were studied for the quantitative chemistry, with the following results:

Variations Per 100 c.c.

Total Nitrogen	20.86 -34.5	mg.
Non-protein Nitrogen	12.82 -17.25	"
Urea	4.54 -14.18	"
Creatinin	0.487- 0.765	"
Creatin	0.563- 0.735	"
Sugar	Trace- 0.60	per cent.

Table 2 (page 238) gives the spinal fluid findings.

The examination of the eye-grounds and the Von Pirquet test may be of help in making the diagnosis.

Tubercles in the choroid may be found, particularly if the meningitis is a part of a miliary tuberculosis. Choked discs are usually present.

According to Holt, "the Von Pirquet gives reliable information except in moribund cases, in those extremely prostrated or with very

TABLE 2.—SPINAL FLUID FINDINGS IN TUBERCULOUS MENINGITIS

Spinal Fluid Findings	Under 2 Years	2-10 Years	Over 10 Years
Amount of fluid withdrawn			
Under 20 c.c.....	29	14	11
20 c.c. to 40 c.c.....	89	89	30
Over 40 c.c.....	56	72	9
Character			
Clear.....	117	135	39
Hazy.....	4	5	2
Slightly hazy.....	20	9	5
Slightly bloody.....	20	11	2
Cytology			
Over 90 per cent. mononuclears.....	73	80	28
75 to 90 per cent. mononuclears.....	59	56	14
50 to 75 per cent. mononuclears.....	15	10	1
50 to 70 per cent. polymorphonuclears.....	5	3	..
70 to 95 per cent. polymorphonuclears.....	2	2	..
Albumin and globulin			
++++.....	55	50	15
+++.....	55	48	21
++.....	48	59	9
Less than ++.....	10	10	2
Fehling's test*			
+++.....	10%
++.....	32%
.....	38%
± or -.....	20%
T. B. bacilli found in smear.....	92	82	23
T. B. demonstrated by animal inoculation.....	52	47	14

* Per cent. on one hundred cases owing to change in recording findings.

poor circulation. A positive reaction was obtained in 161 out of 194 cases tested."

Diagnosis.—Tuberculous meningitis must be diagnosed from a number of conditions, these conditions being somewhat different at different stages of the disease.

In the earliest stages, the fretfulness and irritability of the child may be attributed to teething; or when the vomiting and constipation appear, to digestive troubles, enterocolitis, etc. Even at this stage, the irregularity of the pulse may make one suspicious; and the progressive character of the symptoms and their failure to respond to treatment will strengthen this suspicion. A low irregular fever is usually present. The spinal fluid will be of help in the diagnosis, even if it does not determine it, for the increase in cells, and in albumin and globulin, even if the reduction of Fehling's is normal at this early stage and tubercle bacilli cannot be found, will rule out a meningeal irritation with the gastro-intestinal condition. In cases of meningism, the fluid is normal in character, though increased in amount.

Such a spinal fluid as has just been described will suggest four other possibilities, a brain abscess or tumor (though in tumors the fluid may be normal), a syphilitic meningitis, poliomyelitis and epidemic encephalitis. A brain abscess usually has a history of a primary infection preceding it—brain tumors are rare in young children while tuberculous meningitis is rare in adults—and both these conditions usually give localizing symptoms of a more marked nature than does tuberculous meningitis. Syphilitic meningitis is also uncommon in young children and a Wassermann reaction and response to specific therapy will almost entirely clear the diagnosis.

Poliomyelitis, especially the encephalitic type without paralysis, offers a more difficult problem. While most cases of poliomyelitis have a history of a sudden onset, and most cases of tuberculous meningitis a slow onset, the reverse may occur in either sufficiently often to puzzle the observer a great deal. Especially is this the case when a tuberculous meningitis shows a facial paralysis, and still more if it shows a mono- or a hemiplegia. Of course, the tendency of poliomyelitis is to clear up, and of tuberculous meningitis, to progress, but poliomyelitis may clear up very slowly, and tuberculous meningitis often shows marked temporary improvement.

The most difficult diagnosis of all is from epidemic encephalitis. While certain generalizations may be made they are of not much help in deciding in regard to a particular case. For example, the age distribution shows a far larger proportion of adults and older children suffering from epidemic encephalitis than from tuberculous meningitis. In 274 cases of epidemic encephalitis studied by the author only 36 cases were under two years of age, while of 547 cases of tuberculous meningitis 238 were under 2 years of age. The onset, while usually gradual in both, is somewhat more often abrupt in encephalitis, and in this type of case death may occur within seven to ten days, a shorter duration than tuberculous meningitis usually has. On the other hand fatal cases of encephalitis may die after 2 to 6 months, a longer period than cases of tuberculous meningitis show, although Harbitz reports that in rare instances cases of tuberculous meningitis may run a course of several months. Diplopia is occasionally present in tuberculous meningitis, and frequently in epidemic encephalitis. In both conditions more or less transient paralyses, especially of the cranial nerve, are common and so also are loss of reflexes and dilatation and sluggish reaction of the pupils. Other neurological manifestations that may occur in epidemic encephalitis, such as catatonias, twitchings of localized groups of muscles, muscular tremors, a slurring, hesitating speech, refusal to speak or marked loquacity, profuse sweating, etc., are certainly very unusual in tuberculous meningitis.

Of course, the tuberculin reaction will be of value in young children especially, and the demonstration of tuberculous lesions elsewhere. Tubercles of the choroid will make the diagnosis. But in many cases the examination of the spinal fluid will be the determining factor for both poliomyelitis and epidemic encephalitis. While early in tuberculous meningitis the fluid may show a normal reduction of Fehling's and it may not be possible to demonstrate the tubercle bacilli, later the reduction of Fehling's in almost all cases becomes greatly diminished or absent and the tubercle bacilli should be found by smear in about 75 to 80 per cent of cases. Animal inoculation will also be of value. Of course, in both poliomyelitis and epidemic encephalitis the cell count is usually lower and the increase in albumin and globulin less than in tuberculous meningitis,

indeed in both conditions in rare instances the spinal fluid may depart very little from normal, showing perhaps only a slight increase in the cells or in the protein content.

Epidemic meningitis, while it usually has an abrupt onset and more marked meningeal symptoms than has tuberculous meningitis, sometimes begins rather gradually and runs a slow progressive course, so that clinically, it strongly resembles tuberculous meningitis. If there is a tuberculous family history, the diagnosis is still more difficult. A lumbar puncture will help to clear up the diagnosis but it may have to be repeated before the physician can be sure what disease is present.

The fluid in such a case of epidemic meningitis, if it is tending toward spontaneous recovery, will usually be only slightly cloudy, with a preponderance of polymorphonuclears; the albumin and globulin will be increased, and the reduction of Fehling's will be diminished. It may or may not be possible to demonstrate meningococci. It must be remembered that a somewhat hazy spinal fluid with over 50 per cent. polymorphonuclears may occur in acute cases of tuberculous meningitis. But in these cases the tubercle bacilli are very easy to demonstrate.

So, in this, as in the other diseases which must be differentiated from tuberculous meningitis, the spinal fluid will determine the diagnosis—if not by the first fluid, then by subsequent examinations.

In all these cases, a positive Von Pirquet in young children will have much weight in determining the diagnosis of tuberculous meningitis, and the finding of tubercles in the choroid will render the physician certain.

Treatment.—While no curative measures are known, it is the opinion of most observers that *lumbar puncture* is followed by a temporary amelioration of symptoms and that patients who have frequent lumbar punctures (perhaps every 2-3 days) live somewhat longer than those who do not receive this treatment. Hollis and Pardee recommend the repeated *intraspinal injection of antimeningococcic serum* and report cases recovering after this treatment. An **ice cap** on the head will relieve the headache to some extent, though early in the disease **bromids, chloral**, or small doses of **morphin** may be necessary. The **bowels** should be **kept open**, preferably by **enemata** or **high colon irrigations** after an initial dose of **calomel**. The patient should be given simple and **nourishing food** at frequent intervals. There is no indication for limiting the diet, except according to the patient's digestive ability.

Many attempts have been made to devise some effective treatment. **Surgical measures**, trepanning and draining the cranial cavity, **ventricular puncture**, establishing permanent drainage through the lumbar region, have been tried **without success**. The same **lack of success** has attended the **intraspinal injection** of various substances, **electrargol**, **colloidal radium**, sterilized air, etc. **Tuberculin therapy** has also been tried, **unsuccessfully**.

Prognosis.—Fortunately tuberculous meningitis is not common, for few other diseases are so fatal. It is one of the hardest situations that a physician has to face—seeing a child or a young adult in the earlier stages of a tuberculous meningitis apparently not critically ill, and realizing that in two or three weeks the patient will almost certainly be dead. Reports of cures have been published, but they are comparatively few, and we must recognize that there are chances for occasional error in the diagnosis. However, in careful review of the literature, Hollis and Pardee

a. Harbitz agree that there are perhaps 40 cases of recovery on record in which the diagnosis seems to them well-established.

Harbitz also calls attention to several cases in literature in which recovery took place from an attack clinically tuberculous meningitis. At the death of these cases from other causes several months to many years later autopsy findings revealed lesions indicating a healed tuberculous meningitis. Most of the recoveries reported have been in adults or older children.

Granting that recovery occasionally takes place, one is constrained to feel very hopeless in the presence of a case, since the reports of Holt, Koplik, Meyer and others, who have studied many cases, agree with those of the author in a mortality of 100 per cent.

While tuberculous meningitis is not common in comparison with most diseases, it is, except when an epidemic is prevailing, the commonest form of meningitis. Holt reports that in his hospital experience, 70 per cent. of the cases of acute meningitis were tuberculous. Still reports 174 cases of tuberculous meningitis, 49 of epidemic and 17 of suppurative meningitis. Out of 1258 cases of all forms of meningitis that the author has seen, 547 were tuberculous.

The duration of the cases is variable, and is therefore very difficult to determine. Holt gives the average duration, in children under three years, from the time when vomiting, apathy and slight fever are noted, as $2\frac{1}{2}$ weeks. Meyer states that the duration is, on an average, 17 days in children up to twelve years, 5 days being the shortest and 43 days the longest. Harbitz reports cases from the literature and one of his own running a course of several months. The approximate duration of the author's cases is shown in Table 1.

According to Holt, "death usually takes place from exhaustion in deep coma, or convulsions develop and continue from 12 to 24 hours before death." More cases apparently die in coma than in convulsions.

According to Meyers, about half the cases develop a terminal bronchopneumonia.

Pathology.—Quoting from Councilman, Mallory and Wright, "Tuberculosis of the meninges in general presents a different anatomical picture from tuberculosis elsewhere in the body, in the extent of the acute inflammatory lesions with fibrinopurulent exudation which accompanies the formation of tubercles and tuberculous tissue."

The dura and pia are congested, the latter being thickened and grayish. Adams divides the lesions into two forms, the disseminated miliary and the solitary tubercle. The miliary form is characterized by the formation of small, white or grayish white granules in the pia. They are about the size of the head of a pin and are scattered along the course of the vessels, being usually most numerous about the base of the brain, the circle of Willis, and along the Sylvian fissures. The process frequently spreads to the convexity of the brain. It is usually bilateral, but one side may be affected more than the other. The character of these miliary tubercles is similar to those found in other parts of the body. Sometimes, instead of the scattered tubercles, small clusters of tubercles develop, which in time coalesce to form nodular masses as large as a walnut, according to Councilman, Mallory and Wright. These are located in the pia arachnoid and may go for a long time unsuspected unless they are situated near the motor area.

In connection with the production of the tubercle, there is a serous, suppurulent or fibrinopurulent exudate, which accumulates at the base

of the brain, in the pia arachnoid, in the brain substance and in the ventricles.

The brain shows lesions of the vessels and of the nerve substance. The vessels have a perivascular infiltration, usually of mononuclears and lymphocytes. The lesions of the nerve substance are both parenchymatous and interstitial.

The ventricles are usually distended, and the choroid plexuses and the ependymal lining are thickened and may show tubercles.

The lesions of the spinal meninges are less marked macroscopically, than are those of the brain, but a histological study shows the tubercles and inflammatory reaction. The lesions of the cord substance and of the medulla are also less marked than in the cerebrum.

Tuberculous meningitis rarely if ever exists without the presence of tuberculous lesions elsewhere. According to Holt, the association is as follows in order of frequency: (1) in infants with general or pulmonary tuberculosis; (2) in children from 3 to 12 with tuberculosis of the vertebrae, hip, knee or ankle; (3) at any age, with tuberculosis involving only the tracheal, bronchial or mesenteric lymph-nodes; (4) much less frequently with pulmonary tuberculosis of older children.

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CHAPTER XIV

MENINGEAL HEMORRHAGE AND PACHYMENINGITIS HEMORRHAGICA

By JOSEPHINE B. NEAL, M.D.

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Definition.—Meningeal hemorrhage may be defined as an escape of blood into the spaces between the meninges. This blood may proceed from the blood-vessels of the meninges themselves, from the plexuses formed by the pia mater in the ventricle, or it may proceed from a hemorrhage in the brain itself, bursting through the substance of the brain into the ventricle or subarachnoid space. In this last event, we are dealing with what Froin calls a cerebromeningeal hemorrhage, rather than with a pure meningeal hemorrhage. It is often quite impossible, however, except by autopsy findings, to be entirely sure whether a case is one of pure meningeal hemorrhage, or whether some cerebral hemorrhage is combined with it. Indeed, English and American authors have little to say on the subject of meningeal hemorrhage, except that occurring in the new-born or that due to trauma. The spontaneous or medical form, as it is called by the French, while far from common, constitutes a very interesting group of cases. In this group falls pachymeningitis hemorrhagica. Whether the hemorrhage is secondary to the pachymeningitis or whether the pachymeningitis is secondary to the hemorrhage is not entirely settled.

Etiology.—PREDISPOSING CAUSES.—The causes may be divided into three general groups: (1) Causes operating in the new-born; (2) traumatic causes; (3) pathological conditions.

CAUSES OPERATING IN THE NEW-BORN.—The chief of these causes is dystocia, from maternal or fetal conditions, resulting in a prolonged labor with instrumental delivery. The injury may be due to the molding of the bones, the pressure of the forceps or the prolonged stasis of the intracranial blood-vessels.

In rare instances, intra-uterine disease may be the cause.

Occasionally, spontaneous hemorrhage of the newly born attacks the meninges or brain. According to Holt, this hemorrhagic condition is not a manifestation of hemophilia, nor is it associated in a large per-

centage of cases with under-development, syphilis or sepsis. Antoine Testas holds, on the other hand, that it is frequently due to these last three conditions and that syphilis, alcoholism or lead-poisoning in the parents predisposes children to meningeal hemorrhage soon after birth.

TRAUMATIC CAUSES.—Severe trauma of any kind—a heavy blow, a fall from a height, diving in shallow water—may cause the injury. Diving in shallow water often results in a hemorrhage of the spinal meninges, usually accompanied by hemorrhage of the cord. The trauma may cause a fracture of the skull or spine, or the rupture of the vessels may be due to concussion. In the case of injury to the skull, it must be remembered that the hemorrhage often occurs at a point opposite to the site of the injury.

Among the traumatic causes may be classed also ulceration of the vessels in cases of caries, necrosis or tumors of the bones of the skull or spine.

PATHOLOGICAL CONDITIONS.—Among the pathological conditions are:

(1) *Pathological conditions of the central nervous system:*

Acute:

Purulent meningitis, tuberculous meningitis, encephalitis.

Chronic:

Pachymeningitis hemorrhagica interna, usually due to alcohol, syphilis or lead-poisoning; arteriosclerosis; brain tumors.

Insanity, usually in cases associated with alcoholism or syphilis

(2) *General infection:*

Small-pox, measles, scarlet fever, pneumonia, influenza, tuberculosis, arthritis, septicemia, anthrax.

(3) *Hemorrhagic diatheses:*

Scurvy, purpura, hemophilia, leukemia.

(4) *Chronic disease of other organs:*

Arteriosclerosis, nephritis (including eclampsia and uremia), diseases of the endocardium, pericardium or myocardium.

(5) *External Influences:*

Insolation, most cases of which, according to Aubert, occur from May to September; changes in gases dissolved in blood, manifested in mountain sickness, caisson disease, aviator's sickness.

(6) *Miscellaneous:*

Convulsions in tetanus, strychnin poisoning, whooping-cough, etc.

When we see so many conditions given as the cause of an occurrence, and when we remember that these conditions exist in the great majority of cases without this occurrence taking place, it seems evident that the real cause must lie elsewhere, perhaps in some inherent weak-

of the blood-vessels in the individual cases in which meningeal hemorrhage occurs.

Symptomatology.—**CLINICAL HISTORY.**—(a) *In the New-born.*—Ante Testas and Castaigne and Paiseau state that there are two forms of this pathological state: first, that in which the infant is born apparently dead, without pulse or respiration, and responds but feebly to efforts at resuscitation; second, that in which the evidences of meningeal irritation appear some hours or days after birth. These evidences are convulsions, most often localized in the facial muscles, but sometimes in the hands and feet; and general convulsions, rapidly developing coma, tension of the fontanel, exaggeration of the reflexes and elevation of temperature. Cyanosis usually develops early. Convulsions in the first few days of life are considered almost pathognomonic of meningeal hemorrhage. The diagnosis is made by lumbar puncture.

(b) *In Forms Due to Trauma.*—The clinical history and evidence show severe trauma. The symptoms appear in three stages:

(1) Symptoms of concussion—state of shock, pallor of skin and mucous membrane, shallow breathing, feeble pulse, loss of consciousness.

(2) Lucid interval, in about two-thirds of the cases. This may last several hours or several days.

(3) Symptoms of compression, developing very suddenly and without warning—loss of consciousness, stertorous breathing, slow pulse, inequality of the pupils.

Focal symptoms—convulsive twitchings, paralysis—are present in a large percentage of cases, especially in epidural hemorrhage. In subdural hemorrhages, the presence or absence of hemiplegia depends on the location of the extravasation of blood in relation to the motor area, the extent of the extravasation, etc.

(c) *In Forms Due to Pathological Conditions.*—Froin includes under meningeal hemorrhage a group which he calls *cerebromeningeal hemorrhage*. In these cases, the hemorrhage takes place in the substance of the brain and bursts through, either into the ventricle or into the subarachnoid space, with a resulting bloody spinal fluid. This is really a cerebral hemorrhage, but it is included because the fact that the spinal fluid is bloody would sometimes render the diagnosis from a pure meningeal hemorrhage quite impossible except on the basis of the autopsy findings. Moreover, this classification is suggested by Froin, whose work along this line is certainly second to none.

According to Froin, the symptoms are as follows: The onset is sudden and usually occurs in an arteriosclerotic patient past middle life. Coma or delirium develops, with stertorous respiration. Hemiplegia is almost always present. There is a Babinski reflex on the opposite side from the lesion if the pyramidal tract is involved, and on the same side if the lesion is in the optic tract. Conjugate deviation of the eyes is found in some of the cases. Convulsions are not uncommon. Contractures may be present. Kernig's sign, stiffness of the neck and other meningeal symptoms are rare in this type of case.

The temperature rises slowly and is more or less elevated in the last days of the illness, which is usually of short duration. A type of this form is described by Klippel. The onset is accompanied by signs indicating a cerebral focus, for example, aphasia under the form of word deafness. In the second stage, following at an interval of one or two days, there are generalized contractures and the usual picture of a meningeal lesion.

The purely meningeal hemorrhages may be divided into two types: (1) encysted or circumscribed—*pachymeningitis hemorrhagica interna*—and (2) diffuse.

Pachymeningitis Hemorrhagica Interna.—The symptoms may be divided into two periods: First, the period of inflammation and, second, the period of hemorrhage.

The first period, according to Klippel, is characterized by headache, which is constant, severe and localized. There may be violent exacerbations, with a sensation of constriction and a feeling as if the cranial bones were being crushed, and there are sometimes radiations of the pain. If vomiting occurs, it is not repeated. There is no fever. There may be constipation, but it is not as obstinate as in a meningitis of the pia. There are repeated attacks of vertigo, ringing in the ears, and, sometimes, a stumbling gait.

The sleep is disturbed by pain and nightmares, but there is usually no delirium nor is there, at this period, profound depression. Some authors mention the mental torpor which begins at this time.

The pupils are constricted. There is often nystagmus but not strabismus. In some patients, a hemiplegic paresis may develop.

There is a difference of opinion in regard to convulsions—some writers comment on their absence, others on their presence.

The second period may develop abruptly, if the hemorrhage is sudden in appearance and considerable in quantity; by degrees, if there are repeated small hemorrhages; and gradually, if the hemorrhage takes place slowly. In any event, profound intellectual torpor and somnolence finally appear, usually developing into stupor. A loss of physical power, either generalized or local in the form of a hemiplegia, is also noted. A peculiarity of this hemiplegia is that it is usually found on the same side as the lesion, in contradistinction to the hemiplegia of cerebral hemorrhages, which takes place on the opposite side.

There may be a diminution of sensation but it is not in proportion to the hemiplegia, nor is it permanent. As to the eyes, there may be nystagmus as in the first stage. The early contraction of the pupils gives place to dilatation. There may be more or less amblyopia. But the eye symptom of real importance is papillitis.

The pulse is usually slow. There may be Cheyne-Stokes breathing. The temperature is often a little elevated, perhaps one or two degrees.

As the disease progresses further, the stupor becomes deeper, there are often automatic movements, apoplectiform attacks, pulmonary congestion, convulsions and, finally, coma.

Non-encysted or Diffuse Meningeal Hemorrhage.—The symptoms depend on a variety of circumstances. The hemorrhage may develop apparently out of a clear sky, or it may take place in the course of some acute disease, as an acute meningitis or a pneumonia. In this latter case, the symptomatology will be merged with that of the primary disease.

The onset is very often abrupt, with vomiting and loss of consciousness, though headache may precede. According to Klippel, there are usually two phases, first a short period in which the patient becomes pale and has a tendency to faint without actually losing consciousness or falling; then there is a loss of consciousness, which is not so complete as in cerebral hemorrhage. An examination of the patient reveals the fact that there is not the complete relaxation and flaccidity of the muscles that is found in cerebral hemorrhage. The muscle tone, on the contrary, often seems slightly increased, as shown by the presence of a Kernig, increased reflexes, and a rigidity of the limbs. As a further development of the rigidity (due, in Klippel's opinion, to the compression, which produces a cortical ischemia with a compensatory hyperemia of the subcortical centers) convulsions occur, general or localized, and marked contractures. "It is a classic precept to regard early contractures, in the course even of an apoplexy, as showing a lesion or a complication of the meninges or ventricles."

Sometimes a paralysis, especially a hemiplegia, develops. The pulse, at first normal or slow, gradually becomes accelerated and, in fatal cases, more and more feeble and irregular. Paralyses are comparatively rare in the purely meningeal type, and so is a Babinski. Aphasia or amaurosis may be present, however. There may be Cheyne-Stokes respiration. The unconsciousness may be prolonged and end in death, or it may gradually clear up and recovery take place. In some cases, where there is a rupture of an important artery, death may take place in a very short time—a few hours or less—and the diagnosis may not be made until the autopsy.

In some cases, the clinical picture is that of an acute meningitis—headache, vomiting, Kernig, constipation, stiffness of the neck, delirium, mental torpor or indifference, perhaps without loss of consciousness.

When the meningeal hemorrhage supervenes on an acute meningitis, its onset is indicated by a sudden change for the worse, and usually by a stuporous or comatose condition. Castaigne and Paiseau prefer to designate these as a hemorrhagic meningitis rather than as a meningeal hemorrhage. To the writer this does not seem necessary or desirable, as it really is a meningeal hemorrhage occurring in connection with a meningitis, just as it may occur in connection with a nephritis, and there is no particular reason for giving it a different name.

Some cases show a tendency to recur, either in a short time or after a period of some years. These different forms show a wide variation in severity, but it does not seem well to multiply types.

Hemorrhage of the Spinal Meninges.—The hemorrhage may be either extra- or intradural. Extradural hemorrhage is usually of traumatic

origin, but it may be due to syphilis or to vertebral caries. Severe convulsions (tetanus, eclampsia, etc.) are also given as causes. In traumatic cases, the symptoms are usually submerged in the general symptoms of injury. Otherwise, they are the symptoms of irritation and compression.

Intradural hemorrhages are more uncommon than the preceding. They are due to about the same causes. They may, like hemorrhages of the cerebral meninges, be secondary to a meningitis. The early symptoms are pain and rigidity of the spine and limbs. There may be convulsive movements. A paraplegia usually follows, often localized in the legs, though the arms also may be affected. The paralysis generally progresses for two or three days before it becomes complete. It is ordinarily accompanied by anesthesia.

Retention or incontinence of urine and feces is an important symptom. If a secondary infection does not develop, the blood may be absorbed and recovery eventually take place. Often, however, complications occur—secondary meningitis, or infection by way of the urinary tract, the lungs or skin lesions. If the paralysis is very pronounced, atrophy of the muscles follows, and stiffness of the joints and tendons. The sphincter trouble may persist for a long time. The diagnosis is practically always made by lumbar puncture, the bloody fluid ruling out hematomyelia, but not cerebral meningeal hemorrhage. This can usually be eliminated, however, by the difference in the clinical picture. Vigneras divided the cases into three groups: traumatic, infectious and spontaneous.

LABORATORY FINDINGS.—In all these cases, the diagnosis is made by lumbar puncture. A true hemorrhagic fluid is distinguished from a fluid in which the blood is due to the accidental puncture of a vessel by the fact that, when it is collected in two or three successive tubes, there is a homogeneous color in all the tubes, while in an accidental hemorrhage the fluid soon clears up. Furthermore, in a meningeal hemorrhage, the blood coagulates very slowly, if at all. Most workers claim that it does coagulate unless a meningitis is present.

When centrifuged, the supernatant liquid is pink or, more often, yellowish. If the hemorrhage is very recent, the supernatant fluid may be nearly colorless and become yellow in subsequent punctures.

The amount of blood in a fluid varies greatly, from almost pure blood (especially in traumatic cases and in the new-born) to only a perceptible tinge. According to Klippel, Aubert and others, the amount of blood may be so small as to show only after the fluid is centrifuged, or it may be completely lacking, in cases later found by autopsy to be meningeal hemorrhage. This is explained by the fact that the blood in the cranial cavity has failed to diffuse into the spinal meninges.

When the puncture is made some time after the hemorrhage has taken place, a yellow fluid, showing few or no red blood-cells, may be obtained. Milian gives to this yellow fluid the name "**xanthochromia**." Sometimes this yellow fluid will coagulate spontaneously. To this the name "**xanthochromia with massive coagulation**" or "**the syndrome of**

"men" is applied. This condition has been ascribed to a walled-off hemorrhage, also to tumors situated near the meninges, to the irritation of Pott's disease, etc. The writer has seen it in cases of epidemic meningitis late in both recovering and fatal cases, where the previous lumbar punctures had shown no evidence of a fresh hemorrhage. The writer has also seen it in two cases that were diagnosed at the time as poliomyelitis, but there is a possibility that these may have been meningeal or cerebral hemorrhages. One case recovered and no autopsy could be obtained on the other, which resulted fatally.

The cytological examination shows many red blood-cells in the hemorrhagic fluids and some increase in the white blood-cells. According to Aubert, if the hemorrhage is abundant, there are many eosinophils. Sabrazès and Muratet described large cells, which they called hematophages and which they attributed in particular to hemorrhage of the cerebral meninges. As their name implies, they are large cells, with their protoplasm sometimes filled with red blood-cells. Together with the leukocytes they are supposed to play a part in the process of hemolysis.

The albumin is increased, depending on the extent of the hemorrhage.

The sugar is also increased, it is stated, in these cases.

The bacteriological examination is negative, except, of course, when the hemorrhage is superimposed upon a meningitis.

In some cases of meningeal hemorrhage, a massive albuminuria is found, and sometimes sugar in the urine.

Diagnosis.—In hemorrhage of the new-born and in traumatic hemorrhage, the attending circumstances aid greatly in the diagnosis, which may also be readily confirmed by lumbar puncture.

In meningeal hemorrhage due to pathological conditions the diagnosis is far more difficult, and several possibilities must be considered.

In early cases of encysted hemorrhages, the headache, pupillary contraction, vertigo and mental torpor, suggest also cerebral syphilis and brain tumor. A Wassermann is not of much value, since syphilis is frequently a cause of pachymeningitis. A syphilitic meningitis, however, is usually located at the base of the brain and gives rise to ocular and facial palsies, neuralgia, optic atrophy and deafness.

The differential diagnosis, from cerebral tumors is especially difficult, as the latter show edema of the disc as does also pachymeningitis. It is, however, less marked in the latter condition.

Tuberculous meningitis may need to be differentiated from early pachymeningitis. In each, paralyzes may develop, but the paralyzes of tuberculous meningitis are usually of the face or eye, while those of pachymeningitis are usually in the form of a hemiplegia.

The headache of pachymeningitis is more intense and more localized, vomiting is less frequent and constipation less obstinate than in tuberculous meningitis. Moreover, tuberculous meningitis is most frequently found in young children, while pachymeningitis usually develops in old people, or, less frequently, in earlier adult life.

in the diffuse hemorrhages, with an apoplectic onset, cerebral hemorrhage must be differentiated. The points in the differential diagnosis have already been considered under the discussion of symptoms. In brief, they are the slower onset, the less complete loss of consciousness, the increase of muscle tone with rigidity, convulsions, and early contractures in meningeal hemorrhage. Also, the hemiplegia of meningeal hemorrhage, if it occurs, is usually on the same side as the lesion, while in cerebral hemorrhage, it is usually on the opposite side.

The diagnosis from diabetic, uremic, alcoholic coma and forms due to drugs must also be made. As the urine sometimes shows sugar and albumin in meningeal hemorrhage, also, a urine examination will not be of great value, unless these substances are absent.

In the cases with marked meningeal symptoms, the diagnosis from acute purulent meningitis must be made, and this is usually very difficult. Fever is usually absent or slight in the early stage of meningeal hemorrhage, but one must remember that occasionally a severe type of meningitis may be practically afebrile.

The absence of a hemorrhagic rash proves nothing, as in many cases of epidemic meningitis no rash is present; its presence also has little value, inasmuch as meningeal hemorrhages occasionally occur in cases of purpura and other conditions with a hemorrhage of the skin.

Poliomyelitis or polioencephalitis, the most fulminating cases, may cause some confusion. Indeed, it has seemed reasonable that some degree of meningeal hemorrhage might occur in these cases, if the congestion was especially marked.

In all these cases, the lumbar puncture is the deciding point in the diagnosis. It must be borne in mind, however, that in rare instances the fluid may show an almost complete absence of blood.

Complications.—There are no particular complications associated with meningeal hemorrhage, except as they may arise in the course of the disease which the meningeal hemorrhage is accompanying.

Sequelæ.—Meningeal hemorrhage of the new-born may, according to Antoine Testas, be followed by Little's disease, contracture, paralysis of the ocular muscles, blindness, deafness, jacksonian epilepsy or epilepsy and mental deterioration.

Traumatic meningeal hemorrhage may be followed by one of the purulent forms of meningitis, by a pachymeningitis, by brain abscess or sinus thrombosis.

In meningeal hemorrhage, due to pathological conditions, the chief sequelæ are mental impairment, loss of memory, contractures, paralysis. Many patients show no sequelæ.

The diseases with which meningeal hemorrhage is associated have been indicated at some length under the etiology. The chief are chronic conditions such as syphilis, alcoholism and nephritis, and the acute infections.

Clinical Varieties and Types.—The clinical varieties follow the etiological grouping closely.

MENINGEAL HEMORRHAGE OF THE NEW-BORN.—Antoine Testas divides these cases into two types:

- A. Those in which the infant is born apparently dead;
- B. Those in which the infant is born apparently well but later develops signs of meningeal irritation and compression.

II. **SURGICAL OR TRAUMATIC CASES.**—Here, the hemorrhage may be:

- A. *Cerebral*—of the cerebral meninges.
 - (1) Epidural may arise from:
 - (a) Wounds of the venous sinuses,
 - (b) Wounds of the middle meningeal artery,
 - (c) Wounds of the internal carotid artery, or
 - (d) The diploë of the bones.
 - (2) Subdural.
- B. *Spinal*.
 - (1) Epidural.
 - (2) Subdural.

III. **CASES DUE TO PATHOLOGICAL CAUSES:**

- A. *Cerebromeningeal*—the hemorrhage being primarily in the brain and bursting through into the ventricles or into the sub-arachnoid space, or both.
- B. *Meningeal*.
 - (1) Diffuse.
 - (2) Circumscribed—Pachymeningitis hemorrhagica interna.

Treatment.—**SURGICAL TREATMENT.**—Hemorrhage in the new-born is sometimes treated successfully by **lumbar puncture**. This should be repeated daily or as often as the signs of pressure indicate, until the symptoms have cleared up. Sometimes a single puncture is sufficient. Brady reports a case with recovery in which as much as 60 c.c. of nearly pure blood was withdrawn at a single puncture. Antoine Testas, Castaigne and Paiseau and others recommend lumbar puncture.

Traumatic hemorrhage may require **surgical** interference, if it is the extradural type or if there is a depressed fracture. Lumbar puncture is the easiest way of relieving excessive intracranial pressure.

Meningeal hemorrhage due to pathological conditions should also be treated by lumbar puncture, repeated daily or as indicated by the signs of increased pressure. The author thinks it is advisable to use some care not to decrease the pressure too much in these cases. Aubert, also, warns against evacuating the meningeal cavity too completely. It has seemed to the writer that withdrawing too much fluid might increase the hemorrhage. In several of these cases, the author has given antimeningitis serum. At the first puncture this was administered because the picture was that of a meningitis and the possibility that the hemorrhage might be associated with a meningitis was considered. Moreover, horse serum

has considerable value in hemorrhage. In any event, several cases have responded very well to this treatment.

GENERAL TREATMENT.—The general treatment consists in keeping the patient absolutely quiet. If necessary, sedatives may be given—**bromids**, **chloral**, or if these are insufficient in moderate doses, small doses of **morphin**. An **ice-bag** often relieves the headache to some extent. In comatose cases, some of the diffusible stimulants may be necessary, **camphor** and **ether**, or inhalations of **ammonia**. **Mercury** or **iodids** may be given, since syphilis is probably a frequent cause of meningeal hemorrhage. If a positive Wassermann is obtained, this treatment should, of course, be pushed.

As retention of urine may be present, especially in the comatose forms, the bladder must receive careful attention. Constipation is often a symptom and the **bowels** must be **kept open**. The **diet** at first should be **liquid** and not too abundant. In general, the treatment is about the same as in cerebral hemorrhage.

Prognosis.—The prognosis in all forms of meningeal hemorrhage is grave. In the opinion of all observers, the majority of cases terminate fatally. Most cases of hemorrhage in the new-born terminate fatally, though cases are reported that have recovered after lumbar puncture. Some of these recoveries have been complete. Most untreated cases that recover are followed by some form of paralysis or mental impairment.

The prognosis in traumatic meningeal hemorrhage depends on the extent of the hemorrhage. If the hemorrhage is extensive, death may take place almost immediately. In small hemorrhages, recovery may take place.

In meningeal hemorrhage due to pathological conditions, recoveries are not infrequent, especially in the type with meningeal, rather than cerebral symptoms. Meningeal hemorrhage complicating an epidemic meningitis is unfavorable to recovery, but not necessarily fatal. (Exact figures are of little value in dealing with a condition relatively so rare, so no attempt is made to ascertain them.)

Pathology.—The following discussion of the pathology is taken from Klippel and Adami.

There is little to be said in regard to the hemorrhage of the new-born and traumatic hemorrhage.

In the spontaneous meningeal hemorrhage, due to pathological conditions there are two quite different pictures—that which is shown when the hemorrhage takes place in the false inflammatory membranes, i.e. pachymeningitis hemorrhagica, and that when the hemorrhage escapes freely into the meningeal cavities. In the first form, the lesion is usually at the top of the convexity of the skull, generally near the median line, sometimes on one side, sometimes symmetrically arranged on both sides. The lesion comprises two elements, the false membranes which are developed first, and the encysted hemorrhage or hematoma which is produced by the rupture of the new-formed vessels in the course of the inflammatory process. The membranes are of varying thickness, due to their being

formed in successive layers. Virchow has counted as many as twenty. The process begins with a very thin deposit of fibrin on the inner surface of the dura. This becomes organized by the growth of capillaries. The membranes thus formed are multiplied and superimposed, and bleeding from the capillaries takes place into them. When the membrane has attained a considerable thickness, sufficient bleeding takes place to give rise to a hematoma. It may be formed suddenly or as the result of successive hemorrhages. Several hematomas are usually found. The smaller ones may be absorbed. The larger undergo various transformations into a serous condition, a purulent condition from secondary infections, or calcification.

The hematoma adheres to the pia mater and rests upon the surface of the brain, causing inflammatory reaction and softening. It is most frequently in contact with the parietal lobe, then with the temporal, frontal and occipital in the order given. The lesions may occur at the base of the brain.

This pathogenesis of pachymeningitis is the one most commonly accepted, and it is backed by such excellent authorities as Cruveilhier and Virchow. By some, however, it is claimed that the process is reversed, that the hemorrhage takes place first, and is then encysted by an inflammatory reaction.

While the encysted hemorrhages belong practically always to the dura mater, the diffuse hemorrhage usually comes from the vessels of the pia, from the choroid plexuses (which are a development of the pia), and from the brain itself. In the case of the dura, the hemorrhages are secondary to a meningitis; in the case of the pia, they are primary. In very small hemorrhages, the blood may remain upon the pia mater, or between it and the brain, without breaking through the arachnoid. In the case of any considerable hemorrhage, however, the blood escapes into the space between the subarachnoid and the dura. When the dura is opened, a layer of blood is disclosed. The blood may be fluid or, more often, clotted. The clots may adhere to the dura or to the arachnoid, usually in the regions of the occipital or sphenoidal lobes. Sometimes they are found in the fossae at the base of the brain. The cerebral convolutions may be more or less compressed by them. The amount of blood varies from 50 to 200 grams. The hemorrhage may arise from the veins or the arteries of the pia. Usually a diseased condition of the vessel walls may be noted, the veins being distended and varicose, with soft, friable walls, and the arteries atheromatous.

Hemorrhage into the ventricles may proceed from a rupture of the vessels of the choroid plexus or from a hemorrhage in the brain substance bursting through into the ventricle. A cerebral hemorrhage may also break through into the subarachnoid space.

Historical Summary.—Meningeal hemorrhage has a rather interesting history. The following are, in brief, the important points: The first case recorded is that of Henry II, who was mortally wounded in a journey in 1559. Blood was found between the membranes at autopsy. This case was described by Ambroise Paré. In 1819, Serres separated

cerebral and meningeal hemorrhage. Credit for this is also given to Morgagni. In 1829 Cruveilhier brought forward the theory of the pathogenesis of pachymeningitis interna hemorrhagica, which has already been described under Pathology. Later, Virchow supported this theory. Millian, Chauffard, Widal and Froin have done much to add to the knowledge of the subject since lumbar punctures were described in 1890.

An important contribution to the study of hemorrhage of the new-born was made by Dr. Sarah J. McNutt in 1885. She showed that the hemorrhage in these cases was most pronounced about the fissure of Rolando and that the spastic paralyses were due to this rather than to a hemorrhage about the spinal cord, as had been assumed by Little.

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CHAPTER XV

PURULENT MENINGITIS NOT CAUSED BY THE MENINGOCOCCUS

BY JOSEPHINE B. NEAL, M.D.

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Definition.—Purulent meningitis is an inflammation of the meninges with a purulent spinal fluid. It may be caused by practically any of the pyogenic organisms. The terms septic, suppurative and secondary meningitis are also commonly used.

Etiology.—The principle organisms causing purulent meningitis other than the meningococcus (the only organism causing meningitis in epidemic form and not included in this chapter except for comparison) are the pneumococcus, streptococcus, *Bacillus influenzae* and staphylococcus in the order named. The tubercle bacillus and the treponema pallidum also produce true meningitis but with a clear fluid. Such meningitis is therefore not classed as purulent.

Table I gives the distribution by age and etiology of over 1,200 cases of meningitis. Meningococcic and tuberculous meningitis are included for comparison and general interest.

TABLE I. DISTRIBUTION OF 1,259 CASES OF MENINGITIS
Age—Etiology

Age	Tu- bercle Bacillus	Men- ingo- coccus	Pneu- moco- cus	Stre- pto- coco- cus	Bacillus In- fluenza	Staphy- lococo- cus	Bacillus coli	Total Number of Cases by Age
3 months	4	17	3	8	2	0	1	35
3 6 months	17	46	3	5	6	0	0	77
6 12 months	63	53	10	2	9	2	0	139
1 2 years	154	67	7	6	10	2	1	267
2 3 years	72	31	8	2	4	1	0	118
3 5 years	77	73	4	5	1	2	0	162
5 10 years	84	95	8	9	1	0	1	198
10 20 years	42	95	6	4	1	2	0	150
Over 20 years	34	72	17	8	0	2	0	133
Total	547	549	66	49	34	11	3	1,250

Reports in the literature reveal the bacillus anthracis, various members of the typhoid-colon group—as bacillus lactic aerogenes, bacillus enteritidis, bacillus typhosus, bacillus proteus, the gonococcus, micrococcus catarrhalis, actinomycosis, blastomycetes and other organisms as the causes of cases of meningitis. The author has also seen two cases due to Friedlander's bacillus; one due to a diphtheroid bacillus, the case being secondary to an old otitis media, also one case due to bacillus pyocyaneus.

A number of mixed infections have also occurred in the service of the Meningitis Division of the New York City Department of Health:

Meningococcus and streptococcus	2 cases
Meningococcus and staphylococcus	1 case
Meningococcus and paratyphoid bacillus	1 "
Streptococcus, pneumococcus and staphylococcus	1 "
Bacillus of the colon group, an anaërobic coccus, gram negative cocco-bacillus, a gramophilic cocco-bacillus, and a yeast	1 "

In each of three instances a meningococcic meningitis was followed in a short time by an invasion of another organism, the pneumococcus, streptococcus and colon bacillus, respectively.

Attention is directed to the small number of cases, three (Table I), due to bacillus coli. Barron, collected from the literature 42 cases of meningitis in infants, of which 39 were three months or younger. Of these bacillus coli was the causative organism in 14 instances. In 35 of our patients of three months or less, the bacillus coli was the etiological factor in only one instance.

The distribution by season is given in Table II.

TABLE II. SEASONAL DISTRIBUTION OF CASES OF PURULENT MENINGITIS DUE TO ORGANISMS OTHER THAN THE MENINGOCOCCUS

Organism	Jan., Feb., March	April, May, June	July, August, September	October, November, December	Totals
Pneumococcus . . .	20	22	10	14	66
Streptococcus . . .	16	19	7	7	49
Bacillus influenza . .	2	6	5	21	34
Staphylococcus . . .	1	1	6	3	11
					160

The distribution shows the tendency of pneumococcic and streptococcic meningitis to occur in the winter and spring months, and of influenzal meningitis in the fall and early winter. The number of cases of staphylococcic meningitis is too small to be at all conclusive.

All of these forms of meningitis are often secondary to an infection elsewhere or to an operation on the nose and throat. The paragraph below shows the primary focus, in so far as we were able to locate it clinically. Of course, failure to locate clinically a primary focus by no means rules out its existence. Careful inquiry often reveals a history of an otitis media or a sinus infection dating back some years. In some instances, however, the cases have seemed to be really primary.

Of the 66 cases of pneumococcic meningitis, the evidence of primary infection when ascertained was: Pneumonia in 5 cases; middle ear infection in 11; a history of some trauma in 3, and nasal infection in 2 instances. In one instance a patient with an old endocarditis developed a suppurative choroiditis before the meningitis was diagnosed. It was impossible to decide whether the meningitis was secondary to the choroiditis or whether both were secondary to the heart lesion.

Of the 49 cases of meningitis, streptococcic in origin, the primary infection was: Middle ear trouble in 14 cases, in 4 of which a mastoid operation had been performed; nasal infection in 2 cases; throat infection in 1 case; and trauma in 1 case.

Of the 34 cases of influenzal meningitis a probable primary focus was located in 9 cases; in 6, pneumonia; in 1, clinical influenza; in 1, cough and cold; in 1, otitis media.

Of the 11 cases of staphylococcic meningitis, the probable primary focus in 2 was a pustule on the nose, osteomyelitis in 1, otitis media in 1, pneumonia in 2, bronchitis, in 1, evidence of a primary septicemia in 2 with a hemorrhagic rash and other symptoms.

One case of bacillus coli meningitis followed the enucleation of an eye after a period of four months. In the other cases listed, the histories have been negative or indefinite.

Symptomatology.—**CLINICAL HISTORY.**—The value of the previous clinical history has already been discussed. It is important to emphasize the fact that although many cases of purulent meningitis not due to the meningococcus give the history of a primary focus, it by no means follows that because there is a possible primary focus, the meningitis is secondary to it. In other words a meningitis due to the meningococcus is not infrequently found where the existence of an otitis media or a pneumonia has made one fearful of a pneumococcus or streptococcus infection. Unfortunately many physicians do not recognize this fact and take it for granted that a meningitis developing during an attack of pneumonia, for example, is due to the pneumococcus. In this way the correct diagnosis and specific treatment are often delayed.

PERIOD OF INCUBATION.—It is quite impossible to make any definite statement in regard to the period of incubation. The time that elapses between the beginning of the symptoms of the primary focus and the development of the meningitis varies from a few days to several years. In the cases where no primary focus is located clinically, there is no guide to determine the date of infection.

MODE OF ONSET.—The onset is variable and is often masked by the symptoms of the primary infection. Although the onset is usually sudden, it varies greatly in its severity. The early symptoms may be somewhat mild—headache, vomiting, moderate fever and perhaps a chill, slight to moderate stiffness of the neck and positive Kernig's sign, clear mentality. Or the onset may be very severe—chill and high fever, persistent vomiting, marked stiffness of the neck and Kernig's sign, and early delirium rapidly passing into coma.

SYMPTOMS DURING THE PROGRESS OF DISEASE.—The symptoms in over 100 cases as we have observed them are charted in Table III. Additional cases seen since this table was prepared bring out no new points. It will be noted that vomiting is a fairly constant symptom. Convulsions are also common, especially in children under two years of age. Stupor is found much more frequently than delirium. In a few cases the mentality is clear well up to the end. While the pupils show reaction to light in a fairly large number of cases when first seen, it has been observed that this reaction is usually lost as the case progresses. The knee jerks may be increased early in the disease, but they are usually lost toward the end. In a certain number of cases they are unequal. The stiffness of the neck and Kernig's sign tend to be moderate rather than marked. In children under a year, they are usually so slight as to be somewhat hard to obtain definitely. This is true also of epidemic meningitis in young children. In fact the classical signs of meningitis are, as a rule, somewhat more pronounced in the case of epidemic meningitis than in the other purulent forms, but this rule has so many exceptions that it is unwise to rely upon it in making a differential diagnosis.

TABLE III. SYMPTOMS IN PURULENT MENINGITIS

	Under 2 Years	2-10 Years	10-20 Years	Over 20 Years
Onset				
Sudden	41	21	9	1
Slow	4	3	..	1
Indefinite	33	10	4	3
Headache				
+	..	16	10	9
-	..	2	..	1
Indefinite	Indefinite	16	3	5
Vomiting				
+	30	19	9	4
-	13	4	..	7
Indefinite	5	11	4	4
Convulsions				
+	32	7	1	1
-	12	14	6	7
Indefinite	4	13	6	7
Cerebration				
Stupor	24	12	1	9
Delirium	..	3	4	3
Irritability	12	7	3	1
Indefinite	12	9	4	0
Normal	..	3	1	2
Pupils				
Equal	15	13	3	5
Unequal	5	4	..	1
Indefinite	28	17	10	9
Reaction to light	12	10	3	5
No reaction to light	16	10	3	4
Indefinite	17	13	7	6
Nystagmus	3	1
Rigidity of neck				
Slight	19	9	..	1
Moderate	16	14	8	6
Marked	2	6	1	5
Absent	1	1	..	1
Indefinite	10	4	4	2
Kernig's sign				
Slight	20	8	1	3
Moderate	12	13	6	8
Marked	1	2	2	2
Absent	5	1
Indefinite	10	10	4	2
Fever				
Below 100° F	4	1	1	..
100°-103° F	15	7	2	2
103°-105° F	23	18	8	8
Over 105° F	5	3	..	5
Indefinite	1	5	2	..
Pulse				
Regular	13	9	3	7
Irregular	12	6	4	2
Indefinite	23	19	6	6
Rate, Rapid	29	17	5	11
Slow	..	3	1	1
Moderate	5	2	2	..
Indefinite	14	12	5	3
Respiration				
Regular	10	8	2	4
Irregular	14	7	3	3
Cheyne-Stokes	5	1
Indefinite	21	18	8	8
Reflexes				
Increased	7	6	..	3
Normal	14	6	3	3
Diminished or absent	14	11	3	5
Indefinite	11	10	4	3
Unequal	2	1	3	1
Paralysis				
Strabismus	4	3	..	1
Other paralysis	1	1
Macewen's sign	36	23	6	8

The course of purulent meningitis varies greatly also. In some instances, the progress of the disease is very rapid and the patient dies perhaps in forty-eight to seventy-two hours from the onset of the symptoms. The author has never seen a case of meningitis due to organisms other than the meningococcus in which death occurred in twenty-four hours or less as may occasionally happen in epidemic meningitis. In most instances, the progress is slow and the case lasts from four to five days to two or three weeks.

Table IV shows the duration as we have observed it.

TABLE IV. DURATION OF PURULENT MENINGITIS

Time	Pneumococcus	Streptococcus	Bacillus Influenza	Staphylococcus
Under 1 week	40	21	12	2
1-2 weeks	17	17	8	2
2-3 weeks	5	2	3	2
3-4 weeks	1	1	5	1
4-5 weeks	1	1	6	2
Uncertain	2	7	—	2
		(2 cured)	(1 cured)	(1 cured)

The author has been struck on two or three occasions by the very high temperature (107° F., [41.7° C.] or over) that cases of pneumococcal meningitis show a day or two before death. The temperature does not remain at this level, but sometimes reaches it two or three times. In a case of staphylococcal meningitis, a temperature of 108° F., 42.2° C. was reached several hours before death.

In several cases representing all types, marked remissions of the symptoms have occurred, so that we have entertained some hope of a favorable outcome in spite of the generally accepted belief that the prognosis is almost absolutely fatal. The temperature has approached normal, the mental condition has become clear, nourishment has been well taken and the spinal fluid has shown a diminution in the number of organisms and a marked tendency on their part to become intracellular. In this group of cases four patients have recovered—two after infection with the streptococcus, one with the influenza bacillus and one with the staphylococcus. In a fifth case, due to the colon bacillus, the patient recovered as far as the meningitis itself was concerned, but after two and a half months of good health developed a hydrocephalus. In a case due to both the meningococcus and staphylococcus (not included in Table I) recovery occurred.

As recovery in the purulent forms of meningitis is so rare, a brief description of the cases may be of interest.

The first patient, T. F., 26 years of age, developed meningitis in August, 1913, following an otitis media. The streptococcus pyogenes was demonstrated by smear and culture in the third and fourth spinal fluids; the first two fluids were sterile although they were very cloudy. The patient was very seriously ill, with all typical signs of a severe meningitis—stiffness of the neck, Kernig's sign, internal strabismus, sluggish reaction of the pupils to light, delirium, irregular temperature, up to 105.5° F. (40.8° C.) Antistreptococcus serum was administered both intravenously and intraspinally and also streptococcus vaccine subcutaneously. Urotropin was also given. The temperature became normal in about two weeks and the patient was finally discharged at the end of five weeks in very good condition. A neuritis affecting the back and the sciatic nerve developed two months later but this improved under treatment. It was evident both from the clinical signs and from the spinal fluid that this patient suffered from a serious generalized meningitis.

The second case of streptococcic meningitis was of quite a different character. It was in a child, R. T., 5 years of age, and like the preceding this case followed an otitis media. A culture of a hemolyzing streptococcus was obtained from the first two spinal fluids. A third spinal fluid was sterile and the child made a complete recovery. At no time did she have signs of a well developed meningitis. She showed only a slight stiffness of the neck, complained a little of headache and ran an irregular temperature up to 105° F. (40.5° C.) She was perfectly normal mentally, had no change in reflexes and did not seem seriously ill. The spinal fluids were only slightly hazy, showed only a small increase in the protein content and had a practically normal sugar content, as indicated by the reduction of Fehling's solution. In other words, from the clinical symptoms, the development of the case and the examination of the spinal fluid, it seemed to be and to remain a localized meningitis. No intraspinal injections were given. The only treatment of a specific nature was lumbar puncture.

The patient suffering from an influenzal meningitis was a child, P. M., two and a half years of age, who had a mild cold and cough four to six weeks earlier. The influenza bacillus was found in 8 spinal fluids which showed all the characteristics of a well developed meningitis. Antimeningitis serum was injected at first. Later, an autogenous vaccine was given intraspinally and at the last three injections, influenzal serum was also used. Clinically, the case was moderately sick with headache, vomiting and irregular fever. During the illness, deafness developed which has become complete and permanent.

Recovery from a case of pure staphylococcus meningitis took place in a child, J. J., two years of age. The meningitis developed after a pneumonia during which there had been meningeal symptoms for which two lumbar punctures had been done; in the second tap the fluid was normal except for a few red blood-cells due to trauma. After twelve days, during which the child was convalescing normally, signs of meningitis again developed with a temperature of 105° F. (40.5° C.) A cloudy fluid was withdrawn showing a pure culture of staphylococcus albus, together with a great increase in the protein elements and a marked decrease in the reduction of Fehling's solution. At this puncture, antimeningitis serum was injected. The staphylococcus appeared in the five subsequent fluids. An autogenous vaccine was injected intraspinally. The fluids then became sterile and the child made a perfect recovery.

The case of mixed staphylococcus aureus and meningococcus infection showed a culture of both organisms in the first two fluids, but both organisms disappeared promptly with the injection of antimeningitis serum.

The case of meningitis due to the colon bacillus was one of unusual interest. It occurred in a baby L. S., 3 weeks old. The fluid was so purulent that it could not be removed by lumbar puncture. Ventricular punctures were, therefore, resorted to. These were performed daily, the ventricles being used alternately; at each puncture from 50 to 100 c. c. of fluid that was almost pure pus were removed. This fluid was also uniformly sanguineous. At many of the punctures, irrigations with normal salt solution were also used to wash out additional quantities of pus. Intraventricular injections of an autogenous vaccine were given. At first the baby's condition seemed desperate. There were frequent generalized convulsions, profuse vomiting and a weak, rapid, irregular pulse. The clinical condition improved much more rapidly than did the fluid, which remained very purulent (a test tube full being from three-fourths to four-fifths sediment) with the colon bacillus present in the first 24 fluids. After the organisms disappeared, the fluid became clearer and less in amount. Finally lumbar punctures were tried successfully. After being under treatment for two and a half months, the patient made excellent progress. He gained normally in weight and seemed in every way to be in a satisfactory condition. Suddenly a hydrocephalus developed. It was impossible to obtain more than a few drops of fluid from the spine and ventricular puncture was followed by only temporary reduction of pressure. A surgeon in neurology advised against operative procedures. The condition lasted for nearly ten months. The child finally died. This is the only instance in our experience in which a hydrocephalus following a meningitis has developed with a considerable interval of good health intervening. Of course in a certain percentage of children under one year of age, hydrocephalus develops during a meningococcic meningitis, but in our cases it has always developed during or immediately after the acute stages of the disease and has usually terminated fatally in a much shorter time. Two cases, however showed spontaneous cure.

LABORATORY FINDINGS.—The spinal fluid examination is the most important part of the laboratory work. In no other way can these various kinds of meningitis be diagnosed with certainty. It is also of interest to culture the pus obtained from the primary focus, if it can be located. Blood cultures are of interest, but very few results have been reported.

The fluid is usually under increased pressure; the appearance varies from slightly cloudy to very purulent depending upon the stage of the disease and the reaction to the infection, not on the kind of organism. It has sometimes been erroneously assumed that one could get some idea of the organism by the appearance of the fluid. This assumption is founded on the study of too few fluids. An attack is not necessarily less severe because the fluid is only hazy. Sometimes the fluid resembles a broth culture of the organism and has only a slight cellular reaction. Such a case is usually rapidly fatal.

If the patient be seen early the first or even a later spinal fluid may show no organisms by smear or culture. Such a case should be treated as if it were due to the meningococcus until some other organism is demonstrated. Occasionally the following will be observed: An early fluid shows organisms by smear or culture; then, after the administration of serum (antimeningitis serum is given as routine, until another organism is identified) the fluid will be sterile for several punctures, the original organisms reappearing later. This condition is probably due to a temporary walling off of the organisms or an inhibition to their development, due to the action of the serum. The organisms appear practically always in the later fluids, though the author recalls one case in which the first fluid was sterile and later punctures were unsuccessful because of the walling off of the ventricles by a plastic exudate at the base of the brain. The etiology would have remained in doubt had not an autopsy been done which yielded the streptococcus.

The organisms should be studied by both smear and culture. The smear should be stained by the Gram method, not with methylene blue, as the morphology of various cocci is so similar that little reliance can be placed on it in making a diagnosis. Pneumococci, streptococci and staphylococci are found not infrequently in the cells, while meningococci are often extracellular, at least in part. Too much reliance cannot be placed even on a smear stained by the Gram method because the Gram positive organisms sometimes hold the stain but feebly in smears from the spinal fluid. A diagnosis from the smear should be only tentative, the final decision depending upon the results of cultural examination.

It will be seen by referring to Table V that the three main types of pneumococci and the fourth group are all represented in meningitis due to this organism.

An interesting point in regard to the organism in influenzal meningitis was first brought out by Povitzky in 1920—that is, strains from cases of influenzal meningitis tend to belong to the same type as shown by agglutination. In 11 strains she has found 6 strains belonging to one type. This is in contradiction to the findings among strains isolated from the respiratory tract during the epidemic of influenza (so-called) which showed only a slight tendency to fall into groups when studied by Valentine and Cooper. Povitzky's observation was corroborated by others who found two main groups in spinal fluids: Group I of 8 strains,

Group II of 3 strains, and one intermediate and three heterogeneous strains.

The spinal fluid shows the characteristics of an inflammatory exudate—the cells are increased with a preponderance of polymorphonuclears. It has already been stated that some of the most rapidly fatal cases show only a slightly cellular reaction; the albumin and globulin are increased in varying degrees; the reduction of Fehling's solution is decreased or absent except when the fluid is withdrawn very early; then it may be nearly normal at the first puncture and decrease or disappear later.

Table V gives certain data in regard to the fluids from over 100 cases. The fluids examined in cases seen later bring out no additional points. The first fluid is the one referred to unless it is otherwise stated.

TABLE V. SPINAL FLUID EXAMINATION

	Pneumococcus	Streptococcus	Influenza B.	Staphylococcus
Amount of fluid withdrawn				
Under 10 c.c.	4	2	3	..
10-20 c.c.	13	10	5	..
20-40 c.c.	17	18	13	4
Over 40 c.c.	12	5	2	2
Character				
Slightly cloudy	6	7	3	2
Cloudy	20	20	11	2
Purulent	9	9	7	2
Bloody	1	1	2	..
Cytology				
No information	1	1	1	..
Predominance of polymorphonuclears	40	30	21	6
Few cells	5	2	1	..
Albumin = Globulin				
+++	20	24	16	4
++	10	4	4	..
+	7	8	3	2
Fehling's test				
+++	1	1	..	1
++	2	2
+	10	..	8	3
-	32	25	15	2
Evidence of improvement				
Cells	..	5	2	2
Organisms	..	5	3	1
Fehling's test	1	..
No. of punctures performed on various cases, if more than one	2, 4, 5, 2, 2	4, 2, 6, 2, 2, 4, 3, 6, 2, 2, 2, 0	2, 3, 10, 19, 4	2, 17
No. of cases having the first fluid sterile	1	2	2	..

A certain number of pneumococcus fluids have been typed. They are distributed as follows:

Type I	11 cases.
Type II	20 cases.
Type III	8 cases.
Group IV	23 cases.

Diagnosis.—Cases of purulent meningitis are to be differentiated from each other and also from epidemic meningitis, from meningism especially with pneumonia and otitis media, meningitis sympathetica with brain abscess and from other conditions.

The differential diagnosis of the various types of purulent meningitis, including epidemic meningitis, can be made only by the identification of the organisms producing them. The symptoms, physical signs and history give information that may easily be misleading. Cases with a marked hemorrhagic rash are more likely to be of the meningococcic type, but a hemorrhagic rash also may occur in other forms of

meningitis if a bacteremia or septicemia be present. The author has observed a hemorrhagic rash in influenzal and staphylococcic meningitis. Herpes, also, is more often found in the epidemic than in the other types. In most instances there is not so much retraction of the neck, opisthotonus nor such a well marked Kernig's sign in other forms of purulent meningitis as in the meningococcic form. This fact may be due in part to the patient's not living long enough to develop these symptoms. The history of pneumonia, otitis media or other possible primary focus should in no way make one certain that the existing meningitis is secondary to it.

Meningism, as the author uses the term, is that condition in which meningeal symptoms arise in the course of some disease with the spinal fluid increased in amount but practically normal in character. The symptoms may be quite as acute as in a true meningitis. Some cases show ptosis or strabismus, failure of the pupils to react to light and loss of knee jerks. These symptoms usually subside after a puncture is done, so the operation seems indicated from both the therapeutic and the diagnostic standpoint.

The differentiation from meningitis sympathetica (which is discussed more at length at the end of this chapter) is also made by the examination of the spinal fluid. Meningitis sympathetica is a reaction of the meninges due to an inflammation in the immediate vicinity without, however, invasion by the organisms. The fluid is usually of a varying degree of turbidity, but it is sterile and shows a normal reduction of Fehling's solution, while the fluid of a purulent meningitis, if not at the first puncture, then later, shows the presence of the exciting micro-organisms and a diminished or absent reduction of Fehling's. In the case of brain abscess meningitis sympathetica and its characteristic fluid is present, but there are usually localizing symptoms also to aid in the diagnosis.

Complications and Sequelæ.—The complications and sequelæ of diseases that have so high a mortality as do these forms of meningitis are necessarily few. One case of streptococcus meningitis that recovered was followed by neuritis in the back and legs. In the case of influenzal meningitis described above, recovery was followed by complete and permanent deafness. Rivers reported a cured case of his own in which complete nerve deafness followed. He collected from the literature reports of sequelæ in three other cases; there was some degree of paralysis in all three and, in addition, partial blindness in two. The case of meningitis due to the colon bacillus described above was followed after two and a half months by a hydrocephalus.

Treatment.—The subject of treatment is difficult to discuss because of the highly unsatisfactory results which have been obtained in spite of the many methods that have been tried. Of course, the general treatment is the same as that of any severe acute infection and does not require a detailed description. In all cases of meningitis, if the headache and restlessness are severe and do not respond to moderate doses of bromides or bromide and chloral, it is better to give small doses of Magendie's solution or some other opium preparation than to use large doses of other drugs. Sometimes small doses of morphin and hyoscin are very effective. The symptoms must be treated as they arise. Cardiac stimulation may be necessary and the nature of the stimulant will be determined by the character of the symptoms.

The methods used for the specific treatment of these forms of meningitis are perhaps best brought out by a brief review of the reports of recoveries and the treatment used.

PNEUMOCOCCIC MENINGITIS.—Lamar collected from the German literature reports of 13 cured cases of pneumococcic meningitis, in 10 of which lumbar punctures alone were used, and in 3, pneumococcic serum was given. Royster reported recovery in two of his own cases of pneumococcic meningitis, lumbar puncture alone being done. A. C. Brown reports a case in which he attributes recovery to the use of a commercial vaccine, administered five times. Two lumbar punctures were done. Cupler reports a case that recovered after four lumbar punctures.

Cummings treated successfully a case with intraspinal injections of a commercial anti-pneumococcus serum. Parkinson reports one case and Savy and Gate report two cases that recovered with lumbar puncture alone. Schilleau and Pasquer report a recovery after the intraspinal injection of electragol, but the nature of the organism was not very satisfactorily demonstrated. Steinforth reports recovery in a case treated with three lumbar punctures and two injections of antimeningitis serum. Sanders reports recovery in a case secondary to a bronchopneumonia, with Gram positive cocci resembling the pneumococci; it was treated with autogenous serum. Of these 24 recoveries it will be seen that lumbar puncture alone was used in 16 instances; in 4, a pneumococcic serum was used; in one, a vaccine; in one, antimeningitis serum; and in one electragol intraspinally, and in one, autogenous serum.

STREPTOCOCCUS MENINGITIS.—Recoveries from streptococcic meningitis are rare. Alexander, Netter, Tedesco, Weaver, Day, McCarthy and MacKenzie report one case each; Leighton and Pringle and the author, two each—a total of 11. In the cases reported by Netter, Tedesco and one of the author's lumbar puncture alone was used. Alexander's case, which was complicated by a purulent labyrinthitis and a subdural abscess, recovered after a radical operation. Both of Leighton's and Pringle's cases were treated by lumbar laminectomy and permanent drainage. Day's case was operated on and drainage established. McCarthy's case was treated with normal human serum intraspinally. Weaver's patient received antimeningococcic serum at first, later, polyvalent antistreptococcic serum both intraspinally and intramuscularly. The author's case of generalized streptococcic meningitis received antimeningitis serum at first intraspinally, later, antistreptococcic serum both intraspinally and intravenously and a polyvalent streptococcus vaccine.

INFLUENZAL MENINGITIS.—In 1921, the author published a study of influenzal meningitis which included a report of 13 recoveries, 12 collected from the literature and one case of her own. In 1922, Rivers published a study of influenzal meningitis containing four additional cases that recovered, one of his own and three collected from the literature. In 11 instances lumbar puncture alone was used; in one, electragol was given intravenously daily for a week; in one, convalescent serum from a case of influenza was given intraspinally and later was combined with subcutaneous injections; in one, vaccine (at first stock and later autogenous) was given intraspinally; later, the vaccine was combined with anti-influenzal serum, in one, an autogenous vaccine was given subcutaneously for three weeks and mercury inunction:

used in the back of the neck for one week in addition to 3 lumbar punctures.

STAPHYLOCOCCUS MENINGITIS.—Meningitis due to the staphylococcus is comparatively rare, as a study of Table I will show. The reports of recoveries are few. Bourges reported a case due to the staphylococcus; he attributed the recovery of this patient to an artificial abscess in the thigh produced by the injection of 1 c.c. of sterile turpentine; several lumbar punctures were also done. Lortat-Jacob and M. Grivot reported two cases cured. In one, a mastoid operation was performed in addition to lumbar puncture and the use of vaccine. In the second, in addition to lumbar puncture and vaccine treatment, a **decompression operation** was performed. They refer to the report of a case treated by staphylococcus vaccine intravenously, with recovery. Rocaz reports a case treated successfully with intraspinal injections of **colloidal tin**, 1 c.c. to a dose. One of the author's cases recovered after intraspinal injections of an autogenous vaccine.

A study of the literature reveals the reports of occasional recoveries from various forms of meningitis due to the pneumobacillus, the paratyphoid group, bacillus pyocyaneus, bacillus coli, etc. One of the author's cases caused by Friedlander's bacillus (pneumobacillus) recovered, and one case due to bacillus coli (already discussed) recovered as far as the meningitis was concerned, but died of hydrocephalus which developed over two months later.

A review of these recoveries leaves one in doubt as to whether the recovery in any case was due to the specific treatment employed in addition to lumbar punctures together with the removal of the foci of infection by operative procedures as in cases of mastoid, etc. Certainly **lumbar punctures repeated daily** are indicated. The relief in symptoms following this procedure is usually fairly well marked. Moreover, there is apparently good ground for believing that in epidemic (meningococcic) meningitis the mortality is lower when lumbar puncture is done regularly than in cases where it is not done. The close analogy of other forms of purulent meningitis to epidemic meningitis argues for the intraspinal use of a specific serum when such a serum exists. A study of the recovered cases shows that in a certain number such a serum has been used.

Pneumococcus Type I serum would seem to be especially indicated in Type I pneumococcus meningitis but there are no reports available as to the results of its use. The author has not been able to give it a satisfactory trial. Lamar gives some evidence of the curative value of a mixture of sodium oleate, immune pneumococcus serum and boric acid. (Each cubic centimeter of the mixture contains 0.1 c.c. of a 1 per cent. aqueous solution of Merck's or Kahlbaum's sodium oleate, 0.2 c.c. of immune antipneumococcus serum and 0.7 c.c. of a 5 per cent. aqueous solution of boric acid). This mixture was injected intraspinally in monkeys in which pneumococcus meningitis has been produced experimentally.

There is considerable difference of opinion in regard to the value of antistreptococcus serum in any form of streptococcus infection, but a study of the recovered cases of streptococcic meningitis shows that this serum was used in two cases.

In 1911, Wollstein published reports of the preparation of her anti-influenzal serum and gave experimental evidence of its therapeutic

value. Two recoveries following its use have been reported by other workers and the author has seen great improvement in several cases thus treated. Influenzal meningitis is too rare for the serum to have had a fair trial. Following the discovery by Povitzky and by Rivers (already mentioned) that the strains of the influenza bacillus from cases of influenzal meningitis tend to fall into groups, a serum has been prepared by the Research Laboratory of the New York City Health Department by immunizing a horse with the prevailing strain. The serum has been used in several cases and in those that were not far advanced when the treatment was begun, there has been a marked improvement both in the clinical condition and the findings of the spinal fluid. However, none of the cases recovered.

In studying the treatment of the cases of purulent meningitis which have recovered, it will be noted that vaccines have been used in a number of instances. For the past few years the author has been interested in the possible therapeutic value of injecting vaccines intraspinally in cases of purulent meningitis. Because the vaccine thus administered is absorbed gradually into the blood stream, there is the hope of getting a reaction intermediate between the rather severe shock when vaccine is given intravenously* and the slower action following the subcutaneous injection. The vaccine is given in serum diluted if a suitable serum is available, or in normal saline, after the spinal canal has been drained as thoroughly as possible by lumbar puncture. If the fluid is very thick, it is sometimes desirable to irrigate the canal. The vaccines used have been stock vaccines of the same variety as the organism or an autogenous vaccine when such could be prepared, as it was desired to get any specific effects which might be obtained in addition to the general protein reaction. The dosage is about the same as for subcutaneous administration and no severe reactions have been observed although we have sometimes worked up to large doses. Under this treatment there has been recovery in one case of staphylococcic meningitis, one case of influenzal meningitis and in one case of bacillus coli meningitis, but in the last case hydrocephalus developed three months later. All of these cases have been described. In addition, there has occurred in several other cases marked temporary improvement both in the clinical condition of the patient and in the spinal fluid which showed a marked decrease in the number of organisms and a tendency for them to become intracellular, as well as an improvement in the sugar content.

A further indication of the possible value of this method of treatment was furnished by a case of epidemic (meningococcic) meningitis. The patient had failed to respond to ten doses of serum and was still showing organisms in the fluid, but after two doses of an autogenous vaccine combined with serum intraspinally, she promptly recovered. While the author does not maintain that the recovery in any of these cases was due to intraspinal administration of vaccine, it would seem that this method of treatment is worth trying in these very unpromising cases.

If the spinal fluid becomes so thick that it cannot be withdrawn by lumbar puncture or if there are adhesions along the spinal canal with a

* An account of the use of vaccine intravenously together with a very complete list of references may be found in the Harvey Lectures, 1916-17, pp. 181, "The Influence of Non-specific Substances in Infections" by Jobling.

making dry tap, the fluid may be removed and the serum given by puncture of the cisterna magna as described by Ayer:

The patient is placed on the side as if for lumbar puncture, with neck moderately flexed. Care is taken to maintain alignment of the vertebral column to prevent scoliosis and torsion. After antiseptic preparation of the skin usually including the shaving of a little hair and local anesthetization with procain, the thumb of the left hand is placed on the spine of the axis and the needle inserted in the midline just above the thumb. The needle may be pushed rapidly through the skin, but should then be cautiously and guardedly forced forward and upward in line with the external auditory meatus and glabella until the dura is pierced. If the cisterna be entered at this angle, there is usually a distance of from 2.5 to 3.0 cm. between the dura and medulla; with the needle less oblique in position the distance between the walls of the cisterna becomes progressively less. Therefore, it is good practice to aim a little higher than the auditory meatus and if the needle strikes the occiput, to depress just enough to pass the dura at its uppermost attachment to the foramen magnum. At its entrance the same sudden "give" is felt as in lumbar puncture. The needle employed is a regular lumbar puncture needle, nickeloid, 18 gauge preferred with bevelled stylet, short on sides but not too sharply pointed. There is rather less variation in the depth of the tissue traversed than in the lumbar region, being in an ordinary sized adult from 4 to 5 cm., the greatest distance in the series being 6 cm. and the smallest 3.5 cm. In spite of the simplicity of the technic, it would in the author's opinion be unfair to the patient to perform cisterna puncture without previous experience at the necropsy table." On account of the danger involved in a sudden movement of the patient, it would not seem safe to perform this operation without general anesthesia unless the patient is in profound stupor.

If there is blocking at the base of the brain, ventricular puncture will be necessary. Flexner states:—"The technic of intraventricular injection of the serum is not attended with particular difficulty in young children with open fontanel. The scalp over the anterior fontanel is shaved and rendered surgically aseptic. The needle which is about 8 cm. long is inserted near the lateral angle of the fontanel about 2.5 cm. from the medial line and is gently pushed downward and slightly inward toward the median line to a depth of about 3 cm. when the spinal fluid will usually begin to flow. In older children and adults a trephine opening is made at a point 3 cm. above and behind the external auditory meatus before tapping the ventricles."

OPERATIVE PROCEDURES.—If there is a focus of infection to which the meningitis is secondary, as, for instance, involvement of the mastoid, it is obviously necessary to remove it before there will be any possibility of successfully treating the meningitis. Various other operative procedures have been tried, such as establishing permanent drainage by a laminectomy or by draining the cisterna magna by trephine. The results of these measures have been discouraging.

Prognosis.—The prognosis of cases of purulent meningitis due to organisms other than the meningococcus is bad but not absolutely hopeless. Certainly enough recoveries are reported to justify one in doing everything in his power in working toward a possible recovery.

Pathology.—The pathology of the purulent forms of meningitis,

while in general much the same as that of the epidemic form, present certain points of difference, also the various types differ in some respects from each other. Holt says, "In a general way the anatomical changes resemble those described in cerebrospinal meningitis, with the exception that the marked changes in the brain substance which are usually dependent upon the long course of that disease are wanting. Acute meningitis due to the pneumococcus is characterized by a more abundant exudation of fibrin and pus than is seen in any other variety of meningitis. The lesion may affect the entire brain, but it is especially marked at the convexity and over the anterior lobes. Sometimes it is limited to these regions, the meninges of the base escaping. The exudate may be so abundant as almost to conceal the convolutions. There is usually less distention of the ventricles than in cerebrospinal meningitis. . . . The lesions of influenza meningitis have differed in no essential particular from those described in the pneumococcus variety. . . . The lesions (in meningitis due to the streptococcus and staphylococcus) consist in a widespread general inflammation of the pia with an abundant exudate of pus, but with less fibrin than in the two varieties previously described."

Koplik says, "In the early stages of the disease anatomically there is dryness and opacity of the pia with hyperemia. Later edematous conditions of the pia supervene, with the formation of lymph and fibrin along the sulci and in the tissue of the pia mater and on its surface. Later, the purulent exudate may extend over the surface of the brain, involving not only the base of the brain, but also the spinal cord. In some cases, the exudate does not penetrate the ventricles of the brain; in others, inflammation extends into the ventricles."

Councilman, Mallory and Wright comment on the large amount of fibrin found in the exudate of these forms, especially in the pneumococcus form, and also on the general absence of large cells, inclosing several leukocytes, which are found rather frequently in the exudation in epidemic meningitis. They continue, "The most marked feature in the process in both pneumococcus and streptococcus meningitis was the acute endarteritis. This condition is similar in kind to the vascular lesions which have been described in tuberculosis of the meninges. . . . There were but slight changes in the tissue of the brain."

Hutinel and Voisin describe the changes in much the same manner as the authorities already quoted. They point out that the exudate may be disposed irregularly, especially in cases of otitic origin, where it may be limited practically to the side on which the lesion of the ear existed. They also call attention to the fact that the spinal meninges are always involved to some extent. It is important to keep this in mind, for many, in using the term cerebrospinal meningitis, feel that they are adequately describing meningitis due to the meningococcus, thereby implying that meningitis due to other organisms is not cerebrospinal.

MENINGITIS SYMPATHETICA AND ASEPTIC MENINGITIS.

An account of purulent meningitis would be incomplete if it did not include two somewhat unusual forms without microorganisms, meningitis sympathetica and aseptic meningitis. The former is due to an

inflammation near the meninges and was called meningitis sympathetica by Plaut and Schottmuller. This condition has been thoroughly discussed by Strauss. It is most commonly found as a result of a brain abscess, but it may occur in connection with inflammation of the various sinuses, or with otitis media. There are usually well-marked symptoms of meningitis, and the question arises as to whether one is dealing with a meningism, a purulent meningitis secondary to the primary infection, or with the condition termed meningitis sympathetica. The diagnosis can be made only by lumbar puncture. If the condition is one of meningism, the spinal fluid, though increased in quantity, will be practically normal in its cytology and chemical constituents, and, of course, negative bacteriologically. The spinal fluid in purulent meningitis has already been discussed. In the case of meningitis sympathetica, the spinal fluid is increased in amount, is usually hazy or cloudy, shows an increase in protein and cellular elements, and a normal reduction of Fehlings, and is negative bacteriologically. It would seem to represent an intermediate stage between a meningism due to an inflammation near the meninges, or the middle ear, and a true purulent meningitis secondary to the primary infection, where the organisms have invaded the meninges. Indeed, cases are reported where a purulent meningitis has developed subsequent to a meningitis sympathetica. But it is extremely rare for a meningism to be followed by a meningitis. It may be a question for discussion, therefore, as to whether we should speak of the early stages of a purulent meningitis in which there is a primary focus of infection near the meninges, and where the spinal fluid is sterile for the first one or two punctures, as a meningitis sympathetica subsequently developing into a purulent meningitis. It would seem, however, that this would only invite confusion and that it is better to consider a case in which the organisms are found in the fluid as purulent meningitis, and to reserve the term meningitis sympathetica for those cases which do not go on to a purulent meningitis at all, or only after a period of improvement has taken place.

Aseptic meningitis is a term which has been rather loosely used to designate a meningitis in which the organisms are not found. In the author's opinion, its use should be limited to those cases in which meningeal reaction is due to the irritation of some sterile substance which has been injected intraspinally. If serum is injected intraspinally when the meninges are not actively inflamed, as in tetanus or poliomyelitis, an acute reaction usually follows, with symptoms of meningeal irritation—headache, retraction of the neck, and vomiting—and showing spinal fluid increased in amount, and cloudy, with a marked increase in the albumin and globulin and cells, which are largely polymorphonuclears. The reduction of Fehlings may be masked by the large amount of proteid material which is present. Bacteriologically such fluids are negative. The extent of this reaction seems to vary greatly with the individual. One of the most purulent fluids the author has ever seen was in a case of tetanus following the intraspinal injection of 10 to 15 cc. of tetanus antitoxin. The puncture made at the time of the antitoxin was injected yielded only a few drops of clear fluid. It is well to bear in mind that this reaction exists, otherwise one may be misled into making a diagnosis of a purulent meningitis on finding a cloudy spinal fluid, after a case with meningeal symptoms and a clear spinal fluid has

been injected with antimeningitis or other serum.

Historical Summary.—The term meningitis seems to have been first used by Herpin in the Army in connection with inflammation caused by traumatism of the head. Later, meningeal symptoms were observed clinically and meningitis was established at autopsy in cases of typhoid, puerperal sepsis and pneumonia.

In 1875, Klebs demonstrated, at autopsy, diplococci, morphologically like the pneumococcus, both in the ventricular fluid and in the bronchial secretions. Eberth and Leyden found isolated cocci and diplococci in a case with pneumonia, and in one following otitis. In 1885, in a small epidemic of meningitis in Cologne, Leichtenstern reported diplococci rarely grouped. The following year Sanger found cocci, encapsulated and pathogenic for mice, which fact enabled them to be identified as pneumococci. The same year Frankel and other observers, including Weichselbaum, pointed out the presence of encapsulated cocci in cases of meningitis. In 1887, Netter made a valuable contribution to the subject of purulent meningitis in his memoirs on pneumococcic meningitis, with or without pneumonia. And in that same year Weichselbaum described the *Diplococcus intracellularis meningitidis*, the cause of epidemic cerebrospinal meningitis.

These notes are taken from Hutinel and Voisin.

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CHAPTER XVI

NEURASTHENIA AND PSYCHASTHENIA

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NEURASTHENIA

Synonyms.—Nervous exhaustion, Nervous prostration, Nervosisme, Nervopathie proteiforme, Nerven-erythismus, Neuræmie, etc.

Definition.—The word neurasthenia (*νεῦρον*, nerve; *α*, privative; *σθένος*, strength) was first used by Beard of New York in 1869 to designate a large and decidedly heterogeneous group of disorders, the common characteristics of which were manifestations suggestive of exhaustion with the absence of any disease of organs. Dr. Beard's original papers in various journals were followed by a monograph in 1880 which contained an excellent description of the clinical manifestations. Although various types of functional disorder had been previously described many years before by different authors this book represented the first effort at generalization. That there were included types of cases which we can now recognize as presenting features which justify separation from neurasthenia is not surprising, for even to-day we are

far from a satisfactory and universally accepted definition. Indeed it seems possible that there is a tendency to err in the opposite direction by attempting too fine a discrimination.

In the process of splitting up the original group of Beard, the name neurasthenia has unfortunately been applied to different sub-groups by different authors. Thus we find, for instance, that Kraepelin reserves the title for a group of manifestations bearing many resemblances to the pictures usually collected under this name but which represent definitely the consequences arising from infectious and other exhausting diseases. At the same time under the heading of psychogenic diseases he describes cases "caused by too strenuous or too prolonged, mental or bodily work," included by most authors with neurasthenia, as "nervous exhaustion" (die nervöse Erschöpfung).

The definition is still further complicated by the fact that neurasthenia-like pictures occur in the course of the evolution of certain forms of insanity, especially general paralysis of the insane and dementia praecox. Some of the milder forms of cyclothymic depression also present great similarities which may make diagnosis difficult and which are, unquestionably, often included in this category.

It therefore becomes imperative to arrive at a definition of the disorder which will make clear the particular cases which should be here included. This is not a matter of purely academic interest but has, indeed, the greatest importance in the every-day practice of medicine. For neurasthenia is of very frequent occurrence and is largely dependent for its outcome upon the ability of the physician to recognize and treat it rationally. The failure to make a correct diagnosis and to adopt the proper mode of treatment is responsible for much unnecessary suffering and disability.

Such a definition will not be attempted at the present moment and we shall content ourselves at this point with the tentative acceptance of the commonly adopted view that neurasthenia is a functional nervous disorder characterized by manifestations suggestive of a state of fatigability and irritable weakness which may be observed in any or all of the bodily organ activities and yet without any evidence of disease.

Having for the time accepted this view of the condition, let us consider briefly the meaning of the phrase "functional nervous disorder" and its relation to alterations in structure. The particular form of functional disorder which is to be characterized as neurasthenic will be discussed later under the heading of pathology after the description of the symptomatology.

In no field of medicine has there been so much controversy as in the conception of functional disorders and even to-day there is a very prevalent belief upon the part of the medical profession that disturbances in function are unthinkable without change in structure. Indeed, in all probability to the majority of physicians, the designation "functional" is only a convenient cloak for ignorance as to the underlying structural alteration.

In very large part this attitude is due to the emphasis which, in the

effort to get away from the vague and more or less metaphysical hypotheses of pathology which obtained in ancient times, has been placed upon the study of the changes produced by disease in individual organs and the immense progress which has been made in the recognition and definition of alterations in structure. But the human body is something more than a mere assemblage of organs, each with a more or less set and specific function to perform. These functions are coördinated and harmonized for the accomplishment of a common aim, that of the welfare of the integrated whole—man as a living being. The mechanism for achieving this integration is the nervous system which thus differs from all other systems of the body in that it serves merely to select and combine the functions of other organs towards a purposeful end.

It may be granted that the possibilities of variation in function of any individual organ are very limited and that each, in health, is capable of being modified in only slight degree, mainly quantitative. But the man as a whole is capable of an almost infinite variety of activities by reason of the different combinations of these more elementary functions brought about through his nervous system.

The situation may well be compared with the effects which can be produced by the piano player who employs only a certain number of wires each so constructed as to produce, when struck, a note of a certain pitch. By setting in action these unchangeable units in different simultaneous and successive combinations and by varying the intensity of action of each individual wire and their combinations, there is produced the integration which we know as music.

To carry the simile further it may be pointed out that even with every wire in perfect tune the combinations may be discordant as well as harmonious in their effect upon the listener. In exactly the same way, without disease of organs, the activities of the man as a whole may be out of harmony with his surroundings. It is to such disharmonious activity that the term functional disorder is applied.

Probably it will at once be objected that when discord results, even if the piano wires are in perfect tune (or health) there must be something wrong with the player who, in the illustration, corresponds with the nervous system. But even this is not necessarily true and there are various possibilities for the development of such results, some of which are worthy of closer consideration.

The player may, it is true, have some disease or defect which interferes with his technic. But he may be merely clumsy because he has not had sufficient training or experience. Again the particular sounds he produces may, though discordant to one observer, be harmonious to others. This may be true whether the player is producing oratorio or jazz.

In exactly the same way the combinations of the activities of human organs may be inappropriate because there is some defect or disease of the nervous system which serves to coördinate them (organic disease). They may, however, function poorly in relation to the environment because training and experience in the selection of the proper reactions

have been unsatisfactory, or again because the man has a different set of ideals from those around him. In the last possibility is included the concept that the man may be either behind or in advance of his fellows but in either case he will be different from them or, in other words, his behavior will be relatively abnormal. Many men have been deemed insane or possessed of spirits and have been hopelessly out of harmony with society in various ways because they were actually ahead of their times. This possibility, however, is one not frequently realized and it is true that the reverse is far more usual.

It becomes then necessary next to consider the functions of the man as a unit, the purposes and aims which govern the selection of the particular combinations, simultaneous and successive, of organ activities which make up human behavior. These are no different from those of the single-celled organisms and may be summed up in the statement that they consist of the effort to maintain life which is inherent in all living matter. All living organisms survive only at the expense of their surroundings and this is just as true of man as it is of any other form of life. The key-note to all behavior is therefore the inherent need to survive and reproduce.

The instincts, as they are called, of self and race preservation are in animals possessing a brain, accompanied by certain subjective states which we know as feelings or interests and it may be said in general that their presence is indicative of, and their intensity is proportional to the importance of the reaction called for by the situation to the struggle for the maintenance of life.

With the development of the association system of the brain, the intelligence, the feeling-colored or emotional reactions have become more and more complex, although they still take place through the same effector organs as in animals with much less developed brains. There has been but little modification in the course of the evolution of man, and that chiefly quantitative, in regard to the muscles and glands with which he is equipped to carry out the vastly complex activities of human social life. Man's brain connections, however, enable him to use these organs to greater and more varied effect. Thus, the ability to foresee the consequences of various situations and reactions, brought about by the memory functions of the brain, has rendered the individual able to recognize the importance to the struggle for self and race preservation of many things and happenings in his surroundings to which no attention would have been paid without such ability.

It is thus possible more or less clearly to differentiate between what may be called primary or reflex feelings or interests, those which mean instantaneous and unconsidered reactions provided for in the autonomic parts of the nervous system, and secondary interests which are the resultant of considerations of past experience and future consequences. The latter represent very largely (to what extent yet remains to be determined) the results of individual training and experience, while the former are the consequence of ancestral experience and represent the nerve connections, and hence the combinations of organ activity, with

which the individual is endowed by inheritance and through which, by modification of successive and simultaneous combination, he must express the secondary feelings and interests resulting from his own individual education.

The possibilities in the way of such modifications, as the result of education, are necessarily limited by the structural development of the brain in the form of association pathways which establish the necessary connections between the lower and more reflex nerve centers controlling the receptor and effector organs. Large differences undoubtedly exist between individuals in this respect, and we thus are compelled to take into consideration, when discussing the kinds of reaction which an individual makes to his surroundings (his behavior), both his endowment (heredity) and his training and experience (environment). Under the heading of endowment there are probably many other factors which enter into the determination of the behavior of an individual besides the richness of association pathways in his brain, one of which is the energy or intensity of interest (primary and hence also secondary), the origin and physical substratum of which we know nothing but which corresponds with what is usually described as vital energy—whatever this may be.

Man, in order better to wrest from his surroundings the means for continued existence of himself and race, has developed a social manner of life. While this implies increased efficiency in the struggle for man in general, it brings also with it the necessity for regulating the reactions of the individual in such way as to permit the continued existence of the social group. If all men were blindly to seek to maintain themselves and to reproduce their kind without consideration of the needs and desires of their fellows, the social combination would, of necessity, immediately be disrupted. This is only another way of saying that the development of secondary interests, by educating the individual in the control of his primary or autonomic emotional reactions, must take place if he is to be a member of society.

Hence it may be said that a social mode of life brings with it restrictions upon the instinctive acts of the individual and the establishment of rules of conduct which must necessarily bear most heavily upon those relations between individuals which are of the greatest importance, that is to say, those reactions which are accompanied by the strongest (primary) feelings.

Social regulation obviously cannot destroy the instinctive desires and feelings, for, if it did, the very purposes for which the social union between individuals is formed would be nullified. Social or civilized man must therefore develop interests and reactions for the satisfaction of his primitive needs which will not bring him into conflict with his fellows.

In securing a grasp of the manifestations of functional disorder, it is very essential to realize that primitive needs and feelings are unescapable even though modified in expression by social regulation. Feelings, and the bodily adjustments which are an essential part of them, make up such a large part of functional nervous disorder that unless

this be fully comprehended the whole picture must remain disconnected and mysterious.

The particular ways of making this adjustment which the individual selects, depending, as they do, upon his endowment (including his energy of interest) and his training and experience as a result of the conditions of life in which he happens to be placed, may be more or less successful in enabling him to satisfy the inherent cravings of life and at the same time maintain his social relationship. Failure to accomplish this end, be it slight or serious, is what is described as functional disorder. The form of the disorder varies according to the particular character of the secondary interests which are substituted for the primary reactions, and this in turn depends in part upon the endowment of the individual and in part upon the training he has received.

Functional disorder thus represents the adoption of a mode of adjustment of inherent desires (not necessarily conscious) to social requirements which is unsatisfactory to the individual, to society or, perhaps, to both. Obviously the more restriction is placed upon primitive ways of reacting, that is to say the higher the degree of social organization, the greater the modification of primary feelings each individual must make and the greater the possibility of selecting substitute interests which will be unsatisfactory or out of harmony. Hence it is not surprising to find that functional disorders, almost, if not quite, unknown in lower animals, become increasingly prevalent with the evolution of society.

Again, since the secondary interests make use of the same working tools (receptor and effector organs) as do the primary, one must be prepared to find manifestations of functional nervous disorder in the action of the receptor and effector organs of the body without feeling the need to postulate some disease or defect.

With this preliminary survey of the nature of functional disorder, with one form of which we are dealing, we may turn our attention to a more formal description of its manifestations.

Etiology.—**PREDISPOSING CAUSES.**—Reliable statistics upon the etiology of neurasthenia are not available for several reasons, principal among which is the absence of any very exact and widely accepted definition of the disorders which are to be included under this title. The statements here made therefore are based very largely upon personal experience and views as to definition. In reading them due allowance must be made for the fact that the author excludes from this category the more constitutional types which, if included, as they are by most writers, would result in very materially increasing the influence of heredity and faulty construction.

Sex.—The disorder is somewhat more frequent in females than in males but the difference is not large. This opinion is, however, not universally accepted and von Hössling, for instance, asserts that males are much more frequently affected than females, 604 of 832 consecutive cases observed by him being of the former sex.

Age.—Age, as such, is probably not an important factor, but it will

be found that the incidence falls especially within the years of greatest stress. Thus, in the writer's experience, fully one-fourth of the cases have developed, as nearly as can be determined, between the ages of twenty and thirty, thirty per cent. between thirty and forty and 16 per cent. between forty and sixty. It should be noted here that the exclusion of the constitutional types will necessarily eliminate many functional disorders appearing in childhood and adolescence. The period of climacterium in both sexes would seem to have more influence than that of puberty, probably because this brings with it the realization of approaching limitation of activity and involution.

Heredity.—The influence of *heredity* is especially emphasized by most authors, some neuropathic family taint being alleged in the great majority of cases. While it must be admitted that persons of poor hereditary endowment are probably more liable to develop neurasthenia than are those of better construction, it yet seems logical to assume that the greater the degree of familial taint, the greater the probability that the kind of functional disorder in the offspring will be of the more constitutional and degenerative types. The writer's own cases, in so far as they are usable for this analysis (and it must be admitted at once that no exhaustive investigations into heredity were made), tend to bear this out. Thus of 50 consecutive cases, no history of nervous disease or functional disorder in the family was obtained in 58 per cent. In 34 per cent. "nervousness" in some one or more ancestral or collateral members and in 4 (8 per cent.) definite insanity, twice in an uncle, once in a brother and sister, and once in the mother of the patient, were admitted.

Individual Personality.—The personality of the individual, who is liable to develop neurasthenia, is probably an important factor, but experience in the recent war tends to suggest that the particular type of personality is not one which *a priori* would lead to suspicions of the probability of such development, for many so afflicted had been, before entering military service, successful and eminently useful members of society. Excellent descriptions of the particular features in the make-up of persons who develop neurasthenia have been given by Déjerine and Gauckler and are entirely in keeping with the experience of the author. Such individuals are overly emotional in the sense that they take everything seriously and cannot be indifferent in any matter which interests them at all. They are generous and warm-hearted, form close attachments to family, friends and home and evince a deep sense of responsibility and devotion to duty, often colored by vivid imagination as to the consequences of failures or mishaps. Their feelings are expressed frankly and vividly and they are usually active and energetic, their chief fault lying in the tendency for judgments and acts to be guided more by feelings than by abstract calculation. They thus represent, as a class, some of the most likable and useful citizens of any community, unselfish and vigorous in any activity which appeals to the heart and imagination, though perhaps needing some check upon their sentimentality and imagination. Their own feelings and sensibilities share in the general intensity of interest; they are either very sick or very well;

worry and stress are liable to cause irritability and apprehension out of proportion to the real facts of the situation and throughout there seems to be no intermediate ground.

Social Status.—No social group is exempt, but the disorder is far more frequent among brain than hand workers and also in cities than in smaller towns or country villages. Beard in his original description laid considerable emphasis upon apparently greater incidence in American cities and related this to the hustle and stress of life under the conditions of rapid expansion in a relatively undeveloped country which was yet combined with the social restrictions of a highly developed civilization. It has since, however, been established that neurasthenia is just as frequent in the older communities of Europe where social life is highly organized. Probably the greatly increased responsibilities and difficulties of the immigrant as compared with the native-born bring with them a greater liability to neurasthenia, but upon this point the writer has no facts.

Exciting Causes.—Under this heading are considered a group of conditions which appear to act as the precipitating factors of the actual breakdown. In certain cases they may be the actual cause, but far more frequently they represent simply the last straw that breaks down the resistance to conditions of stress which have been operative for some time.

Overwork.—The influence of overwork in the causation of neurasthenia has been so widely accepted that the two terms have come to be almost regarded as synonymous. But to-day there is a steadily growing recognition of the fact that the real significance of overwork in relation to neurasthenia has not been properly grasped and it seems no exaggeration to assert that overwork as such is quite incapable of giving rise to this condition. Experiment and experience both show that, while work if too prolonged will result in the actual exhaustion of fuel for the production of energy in muscles and gland cells, and at the same time give rise to the pouring into the blood stream of a relatively large amount of waste, and more or less toxic, substances, yet the outcome is simply a state of inaction with or without sleep, and thus the opportunity for recuperation.

The conditions of overwork very often, however, do contain within themselves the really important factor in the development of neurasthenia, namely, emotional stress. Work which must be carried on in spite of feelings of fatigue with their indications of the need for rest, always means work in which the emotional importance is especially great. Only when there is some strong incentive to carry on the efforts in spite of fatigue is there any likelihood of overwork.

Emotional Stress.—Emotional stress is certainly a far more potent, and probably the only important, factor in the causation of this disorder. Many of the manifestations of neurasthenia are in fact but exaggerations and fixations of modes of organ function which normally form part of emotional reactions. This is not the proper place to enter into a description of the various somatic changes which are part of

an emotion. It is essential for our purpose, however, to realize that an emotion is not merely a state of mind, but is far more an adjustment of the body as a whole to meet some condition of importance in the struggle for existence and reproduction. Unfortunately it is only too frequently the custom to stress the conscious side—the feeling—of emotion and to consider the bodily changes, the alteration in muscle tonus, the changes in cardiovascular and respiratory adjustment, the changes in the functional activity of glands and the enormous changes in metabolism, as secondary to this feeling. This view hinges largely upon the fact that we are capable of suppressing much of the outward and visible expression of the emotions. Socialized man is capable of repressing the overt acts and words which represent, for instance, the reaction of anger but, if the situation he has to face is such as to determine primitively such a reaction, his body will inevitably adopt more or less this attitude of preparedness to fight even if he does not actually strike with fists or tongue.

Such body changes, which are physiological and not abnormal in character, constitute an essential part of all forms of emotion even in those which are secondary and not primitive, and it will be necessary to give more detailed attention to them when we come to consider the manifestations of neurasthenia in the various systems of the body.

Under the heading of emotional stress must be included especially conditions of increased responsibility, difficulties in meeting the cost of living, business and financial worries or reverses, changes in habitual modes of life and occupation, demands created by illness of self or family with all the restrictions upon personal pleasures which these may entail, misfortune, neglect and misbehavior on the part of those near and dear, the adjustments required by love and marriage, failures in the achievement of anticipated rewards and accomplishments and finally the sudden shocks of accident and strife which may assail us at any time.

Trauma.—Traumatic neuroses are the subject of a special article (q.v.) and hence but little consideration will be given to the traumatic form of the neurosis with which we are dealing. Suffice it here to emphasize the importance of the emotional element of the trauma and its consequences as the true exciting factor in the production of the group of symptoms included under the title of traumatic neurasthenia.

Infective Fevers.—The relation of infectious diseases to neurasthenia requires much more careful consideration. Like overwork they may appear to be the actual exciting cause of a breakdown of this kind in many cases. In fact a group of manifestations, superficially similar in many respects to some of the main features of neurasthenia, is frequently observed following the intoxication of acute infections, especially if they be at all prolonged.

This fact has unquestionably given rise to much serious misunderstanding of the nature of neurasthenia and the settled conviction expressed by many physicians that all neurasthenia is of toxic origin. Some authors, notably the psychiatrist Kraepelin, even limit the use of

the name, neurasthenia, to conditions of chronic exhaustion following fevers or other toxic or exhausting conditions.

In general it may be asserted that toxins which attack the nerve cells give rise in the first place to hyperirritability which is later followed by diminished excitability and, if sufficiently severe and prolonged, to loss of function and even necrosis. Cells in a state of hyperirritability are more easily exhausted and recuperate more slowly than in health.

The outward expression of such a state of irritable weakness we shall find to be one of the characteristic features of the manifestations of the neurasthenic state and it may be very difficult, in cases with toxic disease as an exciting factor, to define when the mode of reaction ceases to be a direct expression of the condition of intoxication and becomes a functional disorder, to which the author believes the title of neurasthenia should be limited. The situation is very similar to that which obtains in chorea. The characteristic disorders of movement of an attack of acute Sydenham's chorea, unquestionably an infectious disease self-limited in duration, may pass over insensibly into a "habit" chorea in which the infection and its evidences in the form of fever, etc., have disappeared. The disorder in movement has become, as the name implies, a habit. In just exactly the same way the irritable weakness resulting from an intoxication of nerve tissue may outlast the toxic cause and become a functional disorder. Only then is it entitled to the designation of neurasthenia.

The relationship cannot be too strongly emphasized because it gives an insight into the true nature of neurasthenia. Admirable examples of quite identical modes of onset were met with in the recent world war. A very large number of the cases labeled "war neurosis" in the American expeditionary forces were illustrations of this mechanism. But in the place of some specific fever as the precipitating factor there were other exhausting causes, loss of food and sleep and severe muscular effort, combined with intense emotional excitement. As a consequence large numbers of men became actually exhausted but recovered within a few days when given plenty of food, rest and encouragement. Most of them did not have a neurosis at all, but they were, with great wisdom, included with the groups to be cared for by the neuropsychiatrist because they were at least potential neurotics. The foundation was laid and it needed but the operation of factors, already existing, to stabilize and render habitual the manifestations of exhaustion for a true neurasthenia to develop. Failure to recognize this danger and to adopt proper preventive measures resulted in an enormous number of such sufferers in the armies of our allies especially during the earlier years of the war.

In exactly the same way, the irritability and fatigability resulting from an intoxication may persist after the toxic cause and its effects upon the tissues have disappeared if, for instance, this method of meeting the conditions of life, by being too ill and tired to take up responsibilities, brings with it a relief from burdens which appear far more intolerable.

Chronic intoxications with lead, arsenic, morphin, syphilis, etc., may, like the acute intoxications of infection, give rise to conditions of real fatigue and exhaustion which must equally be carefully differentiated from true neurasthenia to which they may also lead.

Exhausting conditions such as starvation, hemorrhage, rapidly recurring pregnancy and prolonged lactation, like the exhaustions of war referred to above, may also form the starting point of a true neurasthenia. Just as with the infections, so the primary state of true exhaustion brought about by such factors may pass over into a neurosis if only the adoption of this method of reaction serves to relieve the patient from conflicts and difficulties which appeal to him as the greater evil of the two.

Symptomatology.—CLINICAL HISTORY.—*Mode of Onset.*—The onset of neurasthenia is usually extremely insidious in spite of the fact that a more or less definite date may be given for the actual beginning of the symptoms. Even when some exciting cause for the onset is alleged, such as the occurrence of an accident, an emotional shock, a period of serious stress or an infectious fever, careful study of the history preceding the breakdown will often reveal that evidences of apprehension and a feeling of inadequacy have antedated the alleged exciting cause by long periods of time. In estimating this it is not sufficient to accept the first assertions of the patient or his friends for they are strongly interested in the establishment of a definitely somatic etiology. The desire to find a material and tangible cause for the subjective complaints is, in fact, an essential part of the disorder itself.

In this connection it is interesting to note that in neurasthenia developing as a war neurosis, a period of "incubation" lasting one or two weeks was almost constantly observed between the exciting cause (explosion of a shell, severe exhaustion, etc.) and the development of the symptoms of neurasthenia. Even then it could be found that the fears of inadequacy for his task, the conflict between the instinctive desire to avoid the responsibilities entailed by the position in which the man found himself on the one hand and the recognition of his social duty on the other, had long antedated the event which apparently excited the attack.

The prodromal uneasiness and uncertainty, which the patient has usually honestly endeavored to overcome and which may have been in existence, though unsuspected by others, for long periods, appear so small and insignificant in comparison with the suffering and distress which make up the condition following the breakdown that they are forgotten or ignored, and it is only when the true significance of the manifestations and their relation to the conflict between the instinctive and the social self becomes clear to the patient that he is able or willing to bring them into the picture.

The fully developed disorder may, thus, appear somewhat acutely following a definite and more or less incapacitating stress, bodily or psychic, or it may develop very gradually and insidiously. It may be considered to have arrived at the time when the patient finally gives

way to his feelings of inadequacy and becomes convinced of the existence of some infirmity instead of continuing to recognize and struggle to meet the difficulty in which he finds himself. His feelings have the power to overcome his judgment and have led to an explanation for the failure to meet the situation which has relieved him of the need for doing so.

There is no rule governing the order of evolution of the symptoms once this break has occurred. The particular manifestations which become the fixed point around which the explanations revolve are more or less accidental and peculiar to the individual patient. The nature of the apparent exciting cause, an exploding shell, or a fall with some more or less trivial injury, the chance observation of another patient, the reading of some article, the remarks of a friend or acquaintance, may serve to crystallize the attention upon some particular feeling and a particular organ or system. In exactly the same way, during the evolution of the picture, chance happenings may serve to change the initial fixation of attention from one point to another, the earlier one being, perhaps, even forgotten in the greater sufferings of the present. It therefore frequently happens that, in giving a history of his trouble, the patient may only recall certain symptoms, at one time very prominent, when they are specifically asked for.

It must also be realized that the *course* is very irregular and variable not only in regard to the particular symptoms which are uppermost at the moment, but also as to the severity from day to day and even from hour to hour. Improvements and exacerbations within quite short spaces of time are frequent, especially during the earlier stages of the disorder and these again are, in all probability, dependent largely upon the environment, the intensity of the demand made upon the patient and the degree of success or failure he experiences. With the progress of the condition, however, the periods of comparative comfort tend to become less and the consciousness of illness more permanent.

This variability of the picture, both as to distribution and intensity of the symptoms, may even be considered as one of the characteristic features of neurasthenia, although it must not be regarded as essential, especially when the disorder has been in existence for some time, as it is a tendency of human behavior in general, including feelings and other modes of reaction, to become habitual.

Subjective Symptoms and Physical Findings.—From what has already been said in regard to the nature and etiology of this condition it will be readily understood that the symptoms may be referred to any organ or system of the body. It therefore becomes necessary in describing the symptomatology, to take up the different systems of the body one at a time. It must, however, be distinctly understood that any or all may be affected in one patient and the localization of the complaint in any one part or organ of the body is not characteristic.

No matter which organ or system is selected by the patient as the main object of his complaints, there are certain features in the symptoms which are common to all.

First, the trouble is essentially subjective and the most careful ex-

an examination will fail to reveal any evidence of disease. (Certain qualifications of this statement will be considered later.)

Secondly, all manifestations suggest a condition of irritable weakness evidenced by hypersensitiveness, the early appearance of fatigue upon even slight exercise of function and the failure to recuperate within the usual limits of rest.

Thirdly, the patient experiences a sense of disability, distress and depression which is out of all proportion to the actual condition of health. It is, indeed, far in excess of the sufferings of persons afflicted with real disease of the organs complained of, many of whom are either strikingly hopeful or quietly resigned to the inevitable. This statement, in fact, only reiterates the emotional nature of the disorder.

Fourthly, there may be a striking disagreement between the degree of disability experienced in some one or more activities and the general appearance of vigor and activity presented by the patient in other respects. Many of them recognize this apparent contradiction and make use of it to emphasize the difficulties of their situation. Nobody realizes how much they suffer nor how ill they are and they seem to be always on the lookout for assertions that their troubles are imaginary. This in itself seems to suggest that these patients appreciate, to some extent, the functional nature of the disturbance though they are at a loss to account in any way other than bodily illness for the sufferings they undoubtedly experience.

In describing the symptoms in detail we will take first a group of uncomfortable feelings and sensations in different parts of the body which may be considered as referred to the cerebrospinal portion of the nervous system. Following this we will take up, *seriatim*, the subjective manifestations and physical findings which may occur in the gastrointestinal, cardiovascular, respiratory and genito-urinary systems. These may be regarded as belonging more especially in the autonomic sphere of the nervous system.

SYMPTOMS REFERABLE TO THE CEREBROSPINAL NERVOUS SYSTEM.—

(1) *Subjective Symptoms*.—Under this heading are included a group of manifestations of great frequency, some of them being almost constant and, at times, constituting the major part of the disability. In describing them we will first take up those relating to the mental state and the projection pathways and will then discuss the relation they bear to emotional reaction.

(a) *Mental State*.—The various complaints made by the patient in reference to the performance of mental operations all bear, especially well marked, the stamp of apparent fatigue and exhaustion. The head is described as feeling empty, it is difficult to think or to give attention to anything, memory is bad, everything seems hazy and confused. The patient feels so tired, restless and irritable, everything worries him, his nerves are on edge and he cannot rest. The slightest sound, the ticking of the clock, the children playing in the house, a bright light or the movement of persons or things around may throw him into a veritable frenzy of irritation ending rapidly in complete exhaustion. Usually then

the patient is more or less ashamed of his outbreak, but a recurrence of the irritation is sufficient to bring about a repetition of the upset which he feels utterly unable to control.

As a matter of fact careful study will demonstrate that he is well oriented and, when he makes the necessary effort, he has a clear grasp of the situation and of the happenings in the world around him. There are no hallucinations nor illusions. His memory is unimpaired, although attention is difficult, and he may complain bitterly of memory defect and may use the feeling of inadequacy thus expressed as evidence of the seriousness and hopelessness of his unhappy lot. He cannot recall where he has put things, the names of people, the duties he should perform and even at times asserts that he cannot remember the dates and mode of onset of his present ailments. The effort to do so is painful in the extreme and exhausts him utterly. The complaint of exhaustion as the result of the attempt may be accompanied by all the outward manifestations of such a state, small, rapid pulse, changes in breathing, sweating and intense lassitude. There is no failure, as a rule, on the part of the patient, spontaneously or when properly requested, to make the effort. But he does so with a settled conviction as to the inevitable consequences and may even show some degree of satisfaction at the justification for his fears and warnings as to the outcome of the experiment, when this exhaustion actually develops.

The stream of thought is connected and coherent and presents no real slowing or distractibility. It should be especially emphasized that the sequence of ideas, the reasoning from the premises accepted by the patient, is logical and, while it may be colored by the particular views of physiology and pathology which he has acquired by reading and observation (he may be informed in great detail concerning symptoms and diseases as the result of frequent visits to physicians and conferences with other patients), yet it does not show any oddities or inexplicable twists.

As regards the content of thought, the main interest centers in the patient's present sufferings and fears for the future. The source and origin of the illness is usually regarded as something outside the patient himself, the result of some period of overwork or undue stress, an attack of influenza, an accident or some other more or less tangible happening, though frequently no cause may be alleged. There is no tendency to self-blame except possibly that he was unwise to attempt so much, should have given up sooner, or should have earlier sought medical assistance. This, however, is not expressed in such manner as to suggest a feeling of responsibility for bringing about the condition by any fault such as is seen so frequently in cases of depressive psychosis. The patient is apprehensive and miserable, but this is a misfortune which has befallen him and not something for which he is to blame.

The sufferings and complaints are intensified by their discussion and can, as a rule, be greatly modified by encouraging explanations and reassurances as to the nature and outcome of the illness or by distraction

of the attention to some other topic in which the patient happens to have retained interest.

Another extremely frequent phenomenon which may be considered here is that of *loss of sleep*. Some degree of insomnia is present in a very large proportion of cases, though increased drowsiness and undue sleep are described as occurring occasionally. The complaints of inability to sleep are probably often exaggerated as patients will state that they have not slept for weeks and even months. Such sleep as is secured is certainly extremely light and the least sound is sufficient to arouse the patient. Often he will lie quite quietly and appear to be sleeping when observed, but will state, when questioned later, that he was awake and noticed the entrance of the nurse though he did not make any movement. It may be impossible to find the patient actually asleep even with the most careful observation.

The brief periods of sleep are often disturbed by sudden starts of the limbs, by dreams, generally of unpleasant character, and sometimes by actual nightmares. On waking, the patient complains that he feels more tired than when he went to bed and is consequently increasingly irritable and hopeless.

It is a fact that the patient often appears less well during the earlier hours of the day and only gradually improves and becomes capable of any exertion towards afternoon. He goes to bed at night with a settled conviction that it is impossible for him to sleep without, and sometimes even with, hypnotics. Consequently he finds himself unable to do so and fully prepared to worry through a weary night with nothing to distract him from brooding over his unhappy fate. Thus he actually tires himself out instead of resting.

(b) *Projection Pathways.*—*Headache*, or perhaps more usually an uncomfortable feeling in the head, is extremely frequent. Often the patient speaks of it as headache or pain but when questioned this is liable to be corrected to a sense of pressure or crushing. It is situated most often on the top of the head "as if there were a weight on it," next in frequency in the frontal region or at the sides of the head, more rarely in the occipital region. It may be described as a feeling of a tight band around the head. It is occasionally unilateral, and may thus suggest migraine, but is far more often bilateral.

Accompanying the pain or feeling of pressure there is frequently tenderness upon palpation of the scalp, sometimes general, at others located in special points which may correspond with the points of exit of the cutaneous nerves. Occasionally this tenderness is not spoken of by the patient and is discovered only on examination of the head.

The headache is usually constant and is increased by any effort at fixation of attention or movement. It may also be described as boring or throbbing. Posture as a rule has no influence upon the intensity of the discomfort, although it may be increased by lying down, and it has no direct relation to nausea or vomiting. It is often worse, as are other symptoms, in the mornings, and improves in the afternoon, although rarely does it disappear altogether.

Distraction of attention will often seem to permit the patient to forget the headache altogether but this is usually followed by the assurance, upon inquiry, that it was present all the time. The writer has known patients assert that they have not been free from headache for ten or fifteen years and yet one finds that in spite of it they may have been able to lead a more or less normal social life much of the time.

Tenderness on pressure over the vertebral spines is frequently present and may be general or localized to certain levels, lower cervical, lumbar or midthoracic. This tenderness, sometimes associated with pain or an uncomfortable aching or burning sensation in the back, is perhaps more frequent in traumatic cases in which it led in the past to the diagnosis of "railway spine," "spinal concussion," etc. It may be so severe as to result in continued bed life, the back supported by pillows, and the avoidance of all movement.

Eye Symptoms.—Hyperesthesia of the retina is quite frequent and has already been referred to as an occasion for irritability. Sometimes it becomes so severe that the patient cannot bear an unshaded window. A patient of the author's, a tall, well-nourished, muscular doctor, screamed and became extremely excited when the writer suggested the need for more light in the room to permit him to be examined. He protested that, even through closed eyelids, daylight was extremely painful and would do him serious harm. The use of the eyes for reading or writing may result in rapid fatigue and often increase in headache, with perhaps pains in the eyeballs.

Ear Symptoms.—The undue sensitiveness to sounds has already been mentioned and may be described as a cochlear hyperesthesia but, like the sensibility to light, it is probably far more psychic than peripheral in character. By this it is intended to suggest that there is no real lowering of the threshold of stimulability but rather an increased feeling of discomfort and annoyance upon the perception of sounds, an exaggerated reaction rather than a hyperesthesia.

In addition, uncomfortable buzzing or throbbing sounds in the ears may be extremely annoying. They, in all probability, correspond with the consciousness of the pulsation of the carotids with which we are most of us familiar as part of the sensations of severe emotion. As a somewhat important negative it may be asserted that such sounds are probably never misinterpreted as illusions or hallucinations. Such errors do occur occasionally in psychasthenia, a functional disorder included by most authors with neurasthenia but as a more constitutional type.

Dizziness or swimming sensation in the head, generally transient and corresponding more with the hot flushes to be described with the cardiovascular symptoms, may be present. It is most often brought about by sudden changes in posture, as from a sitting to a standing position or by bending over to pick something up, though occasionally it seems to be more spontaneous. It is not accompanied by nausea and is not a true vertigo in the sense that it represents a definite feeling of rotation.

Disturbances in the Organs of Smell and Taste.—Manifestations in the special senses of smell and taste do not often take a primary place in the picture but they are frequently met with as concomitants of disturbances elsewhere, especially in connection with gastro-intestinal upsets. The odor and taste of food may be dull and lacking or, perhaps more often, be intensely unpleasant and give rise to nausea and even vomiting. In this respect they really form part of the alterations in appetite which will be considered in connection with gastric symptoms.

Apart from this, variations in the secretions of the nose, increase or diminution, are occasionally observed together with uncomfortable sensations of irritation which may lead to unnecessary therapy and the actual development of inflammatory reactions with a settled conviction of permanent and more or less incapacitating (for certain activities in life) chronic catarrh.

Skin Sensibility.—Various paresthesias, numbness and tingling in the hands and lower extremities, sensations of coldness and swelling and uncomfortable feelings of heat and dryness are not at all infrequent. Objective anesthesia, however, does not occur except in cases where there is a combination with hysterical manifestations. Painful areas with tenderness on pressure are often described but are usually the outcome of a belief or fear of the existence of some disease which is popularly, and sometimes professionally, supposed to be accompanied by such pains.

Muscular System.—One of the most striking and constant manifestations of neurasthenia which has done much to stamp the condition with the name it bears, is the condition of rapid fatigability in the muscles upon even slight exertion. The rapid appearance of this fatigue has been demonstrated by ergograph tracings and in various ways. In many cases, however, it is very obvious that the fatigue is associated, not so much with the use of the muscles, as with the performance of certain special acts, particularly those which are the source of the apprehensions of the patient.

Thus, for instance, the piano player whose chief interest or main means of support depend upon this function may become rapidly exhausted and unable to continue playing but yet be able to use the identical muscles to very great effect and without fatigue for other purposes.

With this fatigability there frequently coexist pains and tenderness in the muscles, cramps and spasmodic twitchings or jerkings of the limbs similar to those we all experience after unduly severe or prolonged exercise. These may be so troublesome at night as to seriously interfere with sleep. Frequently they lead to apprehension of some oncoming paralytic disease and the closest observation of the limbs for further evidence of the dreaded result.

The fatigability of the eye muscles has already been mentioned and this same feature may be observed in any group, including the muscles of speech, mastication and deglutition as well as those of the limbs. Indeed it may be so general and so severe that the patient may become

bedridden, moving neither hand nor foot and maintain this attitude for months and even years.

(2) *Physical Findings*.—From the very nature of the condition as defined above it is obvious that there will be but few objective signs and that the results of examination of organ activity are most generally negative. Nevertheless, there are certain observations bearing upon the fatigability and irritability of function which deserve description. These may be taken up in the same order as that followed in outlining the subjective symptoms.

(a) *Mental State*.—Emphasis has already been laid upon the absence of any evidence of real defect in the association pathways of the brain, the essential feature in the picture being a difficulty of continued attention and the rapid appearance of evidences of fatigue. This last feature has been brought out more or less objectively, by requiring the patient to perform some continuous mental task. Thus, Weygandt has demonstrated that in continued addition of figures there is a progressive diminution of the quality of the work and an absence of the improvement which normally occurs with stimulation and practice.

(b) *Projection Pathways*.—Studies of the fields of vision often show a concentric contraction which may increase very rapidly as the test is continued, with the consequence that the field, at first full, may gradually be reduced almost to central vision. The curve on the chart, if the perimeter be rotated regularly, may thus be more or less spiral in form, the so-called helicoid visual field. The color fields show no distortion in relations.

Bunke has described an interesting series of observations which, if confirmed for neurasthenia as here defined, afford a more or less objective demonstration of the presence of hyperirritability of the optic nerve. Under normal conditions and in the insanities (exclusive of syphilitic disease) the strength of a galvanic current applied to the temple which is necessary to give rise to a sensation of light is from two-thirds to one-fourth of that required to cause a contraction of the pupil. In conditions of nervous exhaustion this relation is said to become altered, phosphenes being produced with a current strength of from one-seventh to one-forty-seventh (average one-tenth) of that required to produce a reflex. He further states that as the condition of exhaustion improves the normal ratio is gradually reestablished.

Examination of the ocular fundi reveals no changes in the disc or retina. Power in the eye muscles initially is good but fatigue is rapidly induced and, in consequence, there is liable to be an oscillation of the eyeballs on extreme deviation which becomes more marked as the effort is prolonged. This oscillation is jerky and irregular and is often spoken of as nystagmoid. Firm closure of the eyelids will also often give rise to a tremulous twitching in the lids of a nature similar to the last.

Ear Findings.—No actual change in the acuity of hearing can be demonstrated though the feeling of discomfort upon stimulation, especially with notes of very high pitch, may be markedly accentuated.

Otoscopic examination reveals no evidence of disease and the Rinne test is negative.

Smell and Taste.—In spite of the complaints of disorder in the sensibility of these organs there is no loss of the ability to discriminate between the various test substances. The mucosa may be reddened and dry or there may be an increased secretion due to the irritation produced by applications made with the object of relieving the discomfort. But this catarrhal change is slight and will rapidly disappear if the source of irritation is removed.

Sensory Skin Disturbances.—Apart from the tenderness of the scalp, over the spines of the vertebræ and other parts of the body already described, there is no objective disturbance of skin sensibility.

Muscular System.—In spite of the term *asthenia* (loss of strength) which is used in the name of the disorder there is no loss of power in the muscles of the body. Considerable tact and persuasion on the part of the examiner may be necessary to induce the patient to do his best, but it will be found that strength is fully proportional to the development of the muscles in spite of the complaint of weakness which is often so prominent. The initial efforts, however, are usually much more effective than subsequent ones unless a long period of rest is allowed. Thus, if the patient be requested to squeeze a dynamometer repeatedly, it will be found that there is a rapid, and more or less progressive, falling off in the degree of pressure registered and an absence of the improvement normally seen with practice. For the same reason prolonged contraction often becomes tremulous, a fact noted above in the movements of deviation of the eyes. Similar irregular variations in strength of contraction can be observed in the protruded tongue, about the mouth and face and in the limbs.

Muscle sense and sense of position are not altered and, though movements may be interfered with by the rapid exhaustion of effort as described above, there is no incoördination. The gait may appear weak and shuffling, especially when the patient knows he is under observation, but there is no ataxy nor other characteristic modification. There may be swaying of the body in the erect position and even a tendency to fall due to the sense of weakness but this is not increased by closing the eyes.

(3) *Special Tests.*—The tendon jerks are most usually somewhat exaggerated in degree and obtained very readily but they may be diminished. The patellar jerk is often accompanied by a very uncomfortable sensation of jarring which is described as running up into the body and which is seldom met with except in neurasthenic persons though it should not be considered in any way as pathognomonic. This same exaggerated sensation may be also observed in connection with the other tendon jerks. No clonus is obtainable. Among the skin reflexes, the plantar reflexes are always of flexor type but the skin stimulation may be accompanied by very unpleasant sensations and defense reactions which may make the reflex difficult to elicit. The cold, clammy condition of the feet may also render these reflexes difficult to obtain. The corneal and pharyngeal reflexes are present.

(4) *Relation of the Symptoms to Emotion.*—It now remains to discuss the relation which these manifestations bear to emotion. Some of them quite obviously represent sensations which belong normally to depressive emotions such as we may all experience and to which we attach no special significance because we know that they will disappear with the passing of the emotion. But there are here also a large group of symptoms, the relation of which to emotion is not by any means so clear. These features are so prominent and bear such obvious similarity to those which appear in conditions of exhaustion and intoxication that they have resulted in the selection of the name which the disorder bears.

Concerning the former group we need here say but little. They are quite obviously exaggerated in relation to the apparent cause, the emotion, and in addition they seem to have become, as it were, separated from it and regarded by the patient as independent sensations to explain which he has developed the belief that they are brought about by some disease process. We shall have occasion to deal more fully with symptoms of this type when describing the disorders belonging in the realm of the autonomic nervous system.

The special features we propose to discuss at this point are the apparent fatigability and the irritable weakness. The researches of Cannon and others establish the fact that violent emotion diminishes fatigue and represents an energizing of the various resources of the body toward meeting situations of especial and primitive importance for the maintenance of life. Yet, as Cannon points out, "in the case of the strong emotions, the effect may be paralyzing until there is a *definite deed to perform*." *

It has already been pointed out that the conditions which lead to the development of neurasthenia are particularly the long drawn out struggles in the conflict between instinctive desire and social regulation. The struggle is thus protracted because the patient finds no way out, there is no definite deed to perform and the emotion may hence have a really paralyzing effect upon energy of action instead of stimulation. The lack of the ability for effective effort and concentration of attention which belong with an attitude of perplexity and doubt are well known.

But this does not explain the rapidity of exhaustion of muscular effort and the failure to recuperate with rest. It has already been pointed out that the evidences of fatigue may appear only in relation to certain particular acts and be absent in others. Every physician who has studied neurasthenics realizes that the concentration of effort and energy of which they are capable, while describing in detail their symptoms and disabilities, will often exhaust the doctor and may leave the patient fresher and more comfortable than when he started. These contradictory results compel one to regard the manifestations as psychic rather than physical. How, then, may they come about?

Déjerine and Gauckler have offered a very instructive and illuminating explanation for some of the manifestations of this kind by pointing out that the acts giving rise to fatigue are no longer performed auto-

* Italics in the original.

mentally as in health. As the result of doubts as to his ability to carry them out, the patient attends closely to every step of the act and thus uses up psychic energy which should have no place in its performance. The patient is applying his efforts much with the same degree of attention as if he were trying to perform the task for the first time and we all know how rapidly one becomes fatigued under such conditions, even though the muscles actually used are still capable of being employed for more accustomed tasks without discomfort. It is probable that the energy applied by the neurasthenic individual is even greater than that of a beginner because of the great importance (hence the emotional coloring) which attaches to success or failure.

But this is not the only mechanism at work in the production of the symptoms of irritable weakness. Another and very important element depends upon the establishment of associative memories which play such a large part in our every-day psychic life. An accidental simultaneity between some thing or happening and the occurrence of painful or other feelings often, as is well known, results in the re-arousing of this, perhaps essentially unrelated, object or event in consciousness whenever the emotional state again appears. In this way the symptoms of some severe or trivial illness, the pains belonging to some injury, the muscular pains and exhaustion of some unduly prolonged and severe muscular effort may become associated with the emotional upset that belongs with a situation to which the individual finds it difficult to adjust himself, simply because they happen to have occurred together. If no satisfactory way out of the difficult situation be found it may well happen that the memory of the discomforts and pains belonging to the associated illness, injury or overwork may continue to recur in consciousness with the persistence of the state of doubt and indecision.

That this mechanism is not a fanciful one can be demonstrated by the actual evolution of symptoms in case histories. It is especially well illustrated in many of the war neuroses. The soldier who doubted his ability to endure, and wished to avoid, the horrors and hardships of military life but yet felt unable to get away from it by reason of a strong sense of his social duty, experienced, during an unusually fatiguing march under depressing circumstances, a real exhaustion, accompanied, as usual, by pains, numbness and a sense of weight in the limbs. The more or less anticipated and desired failure which this implied confirmed many of his previous doubts and he thus came to associate the difficulties in his limbs with his apprehensions. In the future the anxiety and doubts as to his ability to face his military duty carry with them the consciousness of pains and discomforts in his legs. In the neurosis which follows the sensations of fatigue of the lower limbs become the prominent symptom.

Furthermore it must not be forgotten that the semi-starvation and unhygienic mode of life resulting from the neurasthenic state may actually produce a condition of intoxication and exhaustion, the manifestations of which will then be added to and will modify the original picture.

SYMPTOMS REFERABLE TO THE REGION OF THE AUTONOMIC NERVOUS SYSTEM.—The work of Cannon and his pupils establishes in the clearest manner that the vigorous reactions which are represented in consciousness as pain or strong feeling, are activated through the sympathetic system which serves to put into action all the available resources of the body for the accomplishment of some purpose imperative for the defense and maintenance of life. That, on the other hand, the cranial and spinal nerves to the viscera, when they pass to the same organs as the sympathetic group, are always antagonistic and serve the purpose of conservation of energy. Thus the cranial nerve supply to the pupil causes contraction and thus conservation of the retina, the sympathetic dilates it and permits a more accurate observation of the surroundings. The vagus slows the heart beat and provides for more rest, the sympathetic stimulates it to increased activity and also raises blood-pressure. The sympathetic activity represents the individual at war when everything must give way to the urgency of the situation, the vagotonic activity of the body is peace time activity when resources are being built up and energy stored for future use.

Emotion therefore implies more or less mobilization of body forces and concomitant interruption of measures of conservation. Hence it should not be difficult to appreciate that prolonged distress and worry are accompanied by profound alteration in the functions of the body organs leading, if prolonged unduly, to an actual exhaustion, without experiencing the need to assume any disease. So intimately are these manifestations bound up with the emotional state of the patient that in order to avoid duplication, we shall not follow the plan adopted in the last section but shall consider the relationship with emotion while describing the various symptoms.

(1) *Gastro-intestinal System.*—*Subjective Symptoms.*—The work of Pawlow, confirmed by Cannon and others, demonstrated clearly that any form of emotion will prevent the secretion of gastric juice or arrest secretion which is already in progress and that this effect long outlasts the actual state of emotion. This implies not only an absence of digestive power but also the absence of desire for food, or appetite. In addition it is also true that biliary and pancreatic juices remain in abeyance and that the churning movements of the stomach and the peristaltic waves of stomach and intestine absolutely cease. Such effects result from all forms of emotion. Cannon in speaking of the muscular activity of the stomach states that "Even indications of slight anxiety may be attended by complete absence of the churning waves." As a matter of fact we are all familiar with these conclusions from personal experience. Under conditions of worry, anxiety or anger we lose appetite and are liable to suffer from indigestion if we eat in spite of it. Furthermore dry mouth, constipation and occasionally diarrhea are well-known concomitants of states of great excitement. We pay but little attention to these phenomena, as we recognize their normality under the circumstances.

All the above manifestations are frequent symptoms of neurasthenia,

a condition in which we have already postulated the existence of worry and apprehension.

The desire for food is very frequently disturbed or lost and the patient states that he only eats from a sense of duty. This of itself is liable to result in the fixation of attention upon the feelings of discomfort which result from the failure of the stomach and other glands to secrete efficient digestive juices. In consequence the feelings of fullness and distension, which occur normally in slight degree after eating, are exaggerated and are used as an additional reason for not eating. From this to a fear that perhaps there is something wrong with the stomach which may be causing all the distress the patient is experiencing, mentally and otherwise, is a short step very often rendered easy, unfortunately, by the unwise attitude and solicitation of friends and doctors. Remedies and dietetic régimes are instituted under lay or professional advice and the patient is thus confirmed in his apprehensions with corresponding increase in the emotional upset and an accentuation of the symptoms already mentioned.

It not infrequently happens that the discomforts after eating become associated in the mind of the patient with the ingestion of certain articles of food, perhaps accidentally, perhaps because there is a real reason in the especially indigestible nature of the food. Spontaneously, or under advice, the patient then is liable to begin to eliminate such articles from his dietary. At first perhaps some relief is secured, but, since these articles were not the real cause of the discomfort, there is a strong probability that the symptoms will recur and articles will then be gradually cut out until the diet is reduced to milk or some other bland and simple material which is taken in only the smallest quantities. Indeed it is not uncommon to meet with patients who have for months and even years subsisted upon an almost incredibly small quantity of milk or soup.

As a secondary result there are produced the consequences of malnutrition, emaciation, anemia and more or less acidosis which will serve to exaggerate the general bodily disorder by the addition of real intoxication.

The actual form in which the complaints are expressed by the patient vary considerably according to his individual experience and attention. Loss of appetite, heaviness and discomfort in the epigastrium, gas formation, eructation, precordial pains and even vomiting are frequent. The vomitus as a rule is very scanty and consists of little but small quantities of food, though occasionally large portions of a meal may be regurgitated. To these may often be added difficulty in swallowing, due in part to dryness of the throat and esophagus, which is an exaggeration of that which regularly accompanies many emotional reactions, and in part to the absence of desire for food.

The mental attitude of the patient towards these manifestations tends to be one of intense depression with apprehension because he transfers to them the feelings of anxiety and worry which really belong to the situation in which he finds himself. Fears of cancer and other severe and incurable diseases of the digestive system are frequent and the

closest attention is paid to any special feeling or observation which would tend to confirm such a fear. When examined or treated by a physician the patient is continually on the lookout for any chance remark or significant gesture which might serve to support this expectation, which is not only feared but even desired.

Symptoms referable to the lower parts of the digestive tract, arising in exactly similar manner, are also very frequent. Constipation is almost constant, though occasionally it may be temporarily interrupted by transient diarrhea. At times such diarrhea is associated with considerable pain and the passage of large quantities of mucus which may be in the form of casts of the bowel. This symptom resembles the condition of mucous colitis which is, however, not necessarily associated with neurasthenia and is probably evidence of some constitutional deficiency somewhat similar to those, whatever they are, which underlie spasmodic asthma.

Physical Findings.—Examination will most often fail to reveal any evidence of disturbance in the organs. Hyper- and hypo-acidity of the gastric juice may be found and there may be evidence of defective stomach motility. But these findings are inconstant and depend upon the degree of emotional unrest in relation to the ingestion of the food. Such disturbances of the stomach activity may be present at one examination and entirely lacking at the next. Distension of the organs with gas, often a prominent symptom in the patient's account of his troubles, may be found and may result from swallowing of air or perhaps be formed in the intestine as the consequence of stasis.

(2) *Cardiovascular System.*—*Subjective Symptoms.*—The regulation of heart activity and vessel tonus is a complex but very important function of the autonomic nervous system in which the reciprocal antagonism between vagus and sympathetic systems is extremely well marked. As elsewhere, the demands of the purposes to be accomplished through the sympathetic group are prepotent and emotional reaction is accompanied by a profound rearrangement of vascular supply which disregards altogether the constructive needs of the body. We are all familiar with the rapid, vigorous action of the heart, the pounding in the ears or temples, the flushing or pallor of the face, the disturbances in sweat gland secretion and the alterations in the erector muscles of the hair which accompany strong feeling.

Many of these changes are accompanied by sensory impulses which render the individual aware of the disturbance in the ordinary routine operation of the organs in question. Especially is this true of the reactions of fear and depressive forms of emotion and it must be remembered that they take place with perfectly normal organs.

Hence it is not to be wondered at that when the emotional stress has been very severe or very prolonged, as when the adjustments made have proven unsatisfactory, undue significance may be attached to the sensations in connection with the cardiovascular apparatus. Fears that there may be something wrong with this system may readily arise, should attention for any reason be specially focussed upon it, and in

ture would imply maintenance of the upset in the circulation and thus lead to further self-study and apprehension and the establishment of a vicious circle.

Neurasthenic complaints include such features as consciousness of rapid heart action with variations in vessel tonus in various regions leading to sinking sensations, flushes to the face and head, dizziness, throbbing in the ears and alterations in sweating. The close observation of the heart action may lead to sensations of irregularity in the beat often expressed as the heart turns over, the heart stops, etc. Necessarily anything in the surroundings which might give rise to feeling, and such happenings in the irritable, hypersensitive state are very numerous, will cause a marked intensification of the discomfort and the conscious feelings of cardiovascular activity.

Under these circumstances it is not surprising that patients fearing the existence of heart or vessel disease adopt modes of life which will be more or less suitable for the disease feared and they then soon become helpless invalids, afraid to move and fearful of everything which disturbs the peace of the surroundings.

Physical Findings.—The heart action is characterized chiefly by the ease with which the rate is altered. The pulse may be normal or a little rapid when the patient is lying quietly and become markedly accelerated by the least effort or exertion of attention. Thus merely sitting up may increase the rate by 10 or 20 beats and the change from a sitting to an erect position cause a still further and similar increase. Arrhythmias of functional type may be observed; they are not, however, nearly so frequent as the complaints of irregularity. The blood-pressure may be low or high, but is often entirely normal. Apart from the alterations above noted the heart is normal in size and though hemic murmurs may be detected there is no evidence of cardiac disease.

Blood-counts are usually normal but may show the consequences of malnutrition in the form of secondary anemia. At times there is a concentration of the blood due to deficiency in water content which may result in a normal number of erythrocytes in spite of an actual anemia. The white cell count presents no constant or characteristic abnormalities. These findings, however, belong not to the neurasthenia as such but to the disturbance in nutrition and metabolism which may result from it secondarily.

(3) *Respiratory System.—Subjective Symptoms.*—Like the cardiovascular apparatus the respiratory system is also subject to profound alterations under conditions of emotion. The rate and depth of respiration are altered and we become more or less conscious of the extra demands in the form of choking sensations, a stitch in the side or feelings of distress. It is true that these are not marked unless the emotion is intense.

In neurasthenia, manifestations in relation to the respiratory system are not very frequent, although complaints of pains in the chest, difficulty in breathing, choking sensations and irregularities in the rate of respiration are sometimes made. These, if, for any reason, the patient

happens to pay special attention to them, may be the starting point of fears of lung disease, especially tuberculosis or pleurisy.

Physical Findings.—Beyond disturbances in the rate and depth of respiration there are no abnormal findings in the lungs.

(4) *Genito-urinary System.—Subjective Symptoms.*—The kidney functions belong perhaps more closely with the cardiovascular apparatus than with the genital system and the two have been grouped together here merely because of custom. Kidney function is disturbed in emotion concomitantly with alterations in blood-pressure and other circulatory and metabolic changes.

The genital system subserves one of the most important instincts of life and the performance of its function is innervated through the sacral group of nerves. Stimulations of the sympathetic interrupt this function; hence emotion results in lowering of sex appetite and the abolition of potency. Because of the great fundamental and instinctive importance of this function, interferences with its operation are necessarily regarded with deep attention and profound feeling.

In neurasthenia kidney function may be altered as the result of the disturbances in the cardiovascular system, and under some circumstances it may become the center upon which the attention of the patient is focussed. Urine may be excreted in large quantities of low specific gravity or in small amounts with high concentration. In the latter case the sedimentation, which is liable to occur as the result of cooling, is often regarded as evidence of some serious disease. Such observations will tend to increase the closeness with which this function is studied and soon other subjective manifestations may be added, such as discomfort and irritation in the urinary passages, pains in the back over the kidneys, frequency of micturition, etc.

Disturbances in the genital functions are frequent, and it is important to distinguish those which belong to neurasthenia from a group of phobias and obsessions often described as sexual neurasthenia but which really belong in a different category and will be considered under the heading of psychasthenia. While fears as to potency with the inevitable result of partial or even complete impotence may develop, these are secondary to the observation of actual disturbance in function.

In common with other appetites and interests the sexual desire is frequently diminished. At the same time the interference with vagotonic activity may result, in the male, in poorly sustained erection, premature ejaculations and nocturnal emissions which are only too liable to lead to anxious self-observance and apprehension with consequent exaggeration of the manifestations themselves. In the female menstruation often becomes scanty and irregular, a fact which may also bring with it fears of premature age, loss of vitality or disease of the generative organs. Pains and tenderness in the testes or ovaries and along the course of the ducts, pains in the back and uncomfortable bearing down sensations are very frequent.

Physical and Laboratory Findings.—Variations in quantity and specific gravity of the urine have already been mentioned. The constit-

ments of the urine other than water as a rule present but little variation from those of health. Oxaluria, phosphaturia and indicanuria have been especially emphasized and it is said that there is at times an excess of uric acid. The indicanuria, which certainly occurs in some cases, is intelligible as a result of the severe constipation which is usually present. The other elements are certainly not a prominent feature and their meaning, if found, is to be referred to factors other than neurasthenia. Similarly albumin and casts are not present and if found should always lead to the consideration of the presence of some other condition. It is to be remembered in this, as in other physical symptoms, that a neurasthenic reaction may coexist with some organic disease.

The genital organs also are typically healthy, although the concomitance of entirely harmless deviations of the uterus, varicocele, hydrocele and other such conditions is possible and may be one of the factors entering into the fixation of special fears in regard to the health of these organs.

Diagnosis.—The essential features for the diagnosis of neurasthenia consist in the existence of subjective complaints of abnormality in the function of body organs, all of which are compatible with the changes belonging to emotional adjustment, without disease of organs. This subject will be discussed in more detail under the heading of pathology and we shall therefore confine ourselves here to possible sources of error in diagnosis. These may be considered in two categories: (1) the distinction from organ disease; (2) the differentiation from other types of functional nervous disorder (with which have been included for convenience some forms of insanity, the functional nature of which is open to discussion).

(1) **DISTINCTION FROM DISEASE OF ORGANS, INCLUDING THE NERVOUS SYSTEM.**—Firstly, it must be stated that organic disease and functional disorder are not incompatible with one another; that disease may be one of the factors composing the situation to which the patient finds difficulty in adjusting himself and thus may assist in causing emotional disequilibrium and possible functional upset; that structural defect or damage may coexist without causal connection with neurasthenia and yet be made use of in the fixation of symptoms; and, finally, that disorder in function may actually lead to structural damage, as, for instance, when tuberculosis develops as one consequence of malnutrition and faulty hygiene which, in themselves, are purely functional in origin.

Secondly, there is a large group of manifestations of actual fatigue and exhaustion closely resembling, on the surface, those of neurasthenia, but which arise on an entirely different basis, viz., the presence of some exhausting disease. Such examples should certainly be excluded from the category of neurasthenia if the definition to be given is accepted. That they may be difficult to distinguish from true neurasthenia appearing as a mode of reaction to a situation of which disease is a part, is obvious. This difficulty must be met by a detailed study of the manifestations, their mode of development and their relation to the direct effects of the disease upon the tissues.

Among those disease conditions which are liable, more or less constantly, to bring about real fatigue and irritable weakness of tissues must be mentioned especially a large group of intoxications. The toxins may be (1) bacterial: the infectious fevers, syphilis (especially the form of parenchymatous involvement of the nervous system known as paresis or general paralysis of the insane), tuberculosis, malaria, rheumatoid arthritis, etc.; (2) exogenous: morphinism (especially during withdrawal), alcohol, lead, etc.; (3) endogenous or auto-toxins: endocrine disturbances, especially Addison's disease and exophthalmic goiter, diabetes, uremia, Bright's disease, etc. In this group mention should be made especially of one type of central nervous intoxication which has on several occasions presented unusual difficulties in diagnosis. This concerns the earlier stages of the subacute combined degeneration of the cord usually associated with pernicious anemia. As is well known, the blood changes may be very late in their development and under such circumstances the patient may, for many months, present only manifestations of excessive fatigability and subjective disturbances in various organs, without objective signs of any damage to the nervous system. In this, as in the other conditions mentioned, there is a true exhaustion and absence of primary emotional stress.

Besides these definitely intoxicating conditions there is also a group that may be included under the heading of exhaustion conditions, which bear a very close relation to the former and may really be toxic. Among them would be included the effects of prolonged inanition resulting from deficient food supply, the inability to ingest and absorb food as the result of esophageal or pyloric stenosis, gastric ulcer, etc., prolonged lactation, rapidly recurring pregnancy, severe loss of blood as the result of injury or disease and prolonged overwork under emotional stress.

This last cause of exhaustion we have already discovered to have a definite and very close relation to the development of a true neurasthenia. But it must also be borne in mind that it gives rise to a real exhaustion which is not neurasthenia. The relation of overwork to exhaustion on the one hand and to neurasthenia on the other is also true of most of the other intoxicative and exhaustive factors alluded to above and serves admirably to illustrate the distinction which must be drawn between these two states. Real fatigue or exhaustion requires only rest and a proper nutritive supply to the tissues for their relief, although the time needed for this purpose will necessarily be proportional to the damage which has occurred. Neurasthenia, on the other hand, is a disorder in behavior and not a true fatigue or exhaustion and hence the remedy is not merely rest and food, although these may be necessary because of the secondary effects of the disturbance in function, but is essentially education in more appropriate methods of reaction.

(2) DIFFERENTIATION FROM OTHER FORMS OF FUNCTIONAL NERVOUS DISORDER.—The definition of functional nervous disorders given in the beginning of this article (p. 270) endeavored to make clear the view that all are types of behavior adopted by the individual as an effort at social adaptation. But while they have this feature in common, it

is important to distinguish them from one another for the reason that the outcome is very different in different types.

The first distinction which has been made between them concerns the question of the particular realm of nervous activity which seems to be involved in the manifestations. For while all are necessarily and essentially psychogenic, yet the mode of expression may concern either the highest levels of the nervous system, the psychism proper, or the lower levels or projection pathways. To the former group would belong those disorders usually classed as functional psychoses (or insanities) while the latter are spoken of as neuroses or psychoneuroses. This subdivision, while pragmatic because of the differences in the relations with society which result, yet is nevertheless symptomatic and not genetic.

Among the psychoneuroses the most important types are neurasthenia and hysteria. Both express psychic reactions in the form of bodily manifestations. For long there has also been included another type which has been designated as psychasthenia. Here the manifestations are essentially psychic, even although necessarily they are expressed, as are even the functional insanities, through the effector organs, these being the only pathway of communication to the outside world. There is now a tendency to exclude psychasthenia from the psychoneuroses, and to group it rather with the psychoses, although it is not an insanity.

If one compares neurasthenia with hysteria, the only two large groups then remaining in the psychoneuroses, one will be immediately struck with certain points of difference.

One of the first of these points concerns the question as to the kind of person (in which must be included both hereditary endowment and the results of early training, in what proportions is not known) who is liable to develop either of these forms. The personality which predisposes to neurasthenia has already been described. The type of individual who develops a hysterical reaction presents features characterized also by emotional excess but in which the expression tends to be especially explosive and short lived, scattered and more or less ill-balanced. Instead of a deep sense of responsibility and devotion to duty we find a tendency to live in the moment and to be diverted by chance stimulations. The interests are focussed upon the patient's own pleasures, the welfare of others tends to be ignored and there is a marked ability to forget whatever is unpleasant instead of preserving and thus learning by experience.

This last feature becomes especially characteristic of the manifestations of the developed hysterical reaction and is in striking contrast with that of the neurasthenic. The latter, as we have seen, suffers intensely and devotes more and more of his time to the study of his disabilities, while the hysteric tends to accept the disability calmly and to ignore his paralyzed or contractured limb or his anesthesia much, as it has been expressed, as if he had forgotten altogether that he ever had such a limb or sensibility.

The paralysis or other hysterical manifestation represents, as a

symbol, the emotional adjustment the individual has been unable to make and is, as it were, thus put on one side and ignored. To this phenomenon is given the title dissociation because it seems to represent a part of the psychic life of the individual which is split off from the general consciousness. Dissociation or isolation of certain experiences is a very important element in many forms of functional disorder and is seen in its most developed form in dementia præcox.

The absence of dissociation in neurasthenia seems to stamp this type as essentially different from all other forms of functional disorder with the possible exception of the manic-depressive psychoses, if these are truly psychogenic as seems possible at least in some cases. Its presence in hysteria, psychasthenia and dementia præcox serves as an important element in differentiating them. The evidences of dissociation to be sought consist in the appearance of odd and unexplainable (on the basis of the apparent state of emotion) features and it is partly by a search for these and the careful weighing of the meaning of every symptom that a diagnosis can be reached.

It has already been asserted that neurasthenia-like manifestations are quite frequent in the early stages of dementia præcox and that some doubt not infrequently arises as to diagnosis. The resemblance to neurasthenia is however not close, for though there may be marked complaints of bodily illness with severe discomfort in various organs, irritability and apprehension these are usually vague and scattered with many oddities and apparent absurdities which are due to dissociation. Ideas of reference are rarely lacking altogether and hallucinations occur frequently. These, in dementia præcox, are dissociations and do not occur in neurasthenia.

The differentiation from mild cases of depression belonging to the manic-depressive psychosis, often called cyclothymia, is frequently far from easy and undoubtedly many such cases are included with neurasthenia. The difference here lies in the fact that the depression is primary and all the symptoms of which the patient may complain are used as evidence of his feelings. There is also a much greater tendency to self-accusation and the allegation of personal responsibility for the condition rather than a conclusion that the patient is depressed because of the condition of the bodily organs. A study of the history may also reveal a history of past, more or less severe and prolonged attacks of "the blues," perhaps alternating with periods of decided elevation of mood. The distinction here is of special importance because of the danger, which always exists in the depressed cyclothymic individual, of suicide, an eventuality which is not to be anticipated in neurasthenia. A not uncommon fear among neurasthenics concerns the possibility that they are "losing their minds" and will end their days in an institution for the insane. This fear is based upon the feelings of emptiness in the head, difficulty in giving attention and reviving memories. The differentiation from insanity is therefore important in order, with the utmost confidence, to be able to reassure the patient. The foundation of this distinction lies in the fact that there is nowhere any loss of contact

with reality. The neurasthenic patient fully realizes the actual relations between himself and society, his grasp of the surroundings is entirely accurate and his reactions to these surroundings are appropriate even if not well chosen. His behavior is based upon actual disturbances of function in the organs of which he complains.

Finally, it remains to discuss the relation of neurasthenia to psychasthenia which has been purposely left to the last for the reason that psychasthenia has been, and to some extent still is, considered only as a special and perhaps "constitutional" variety of neurasthenia. The most striking feature of the psychasthenic reaction, which undoubtedly is responsible for its inclusion with neurasthenia, consists in the appearance of so-called phobias and obsessions or a feeling of compulsory rumination over certain thoughts, feelings or acts. These are often strikingly at variance with the remaining content of the patient's psychic life, although it is true that they may in time come to dominate a very large part of it, and are often described by the patient as something more or less absurd and contradictory, as if introduced, as it were, from without. These facts at once suggest that they are really dissociations, which, as we have said, do not occur in neurasthenia.

Fears, of more or less fixed form, or phobias, do develop as we have seen in neurasthenia, but they are always logically deduced as the consequence of the disturbance in function which is perceived, and is actually present, in the organs of the body. There is thus no real resemblance between the two conditions at all. The fears of the neurasthenic are open to correction by simple explanation, although it may take much time and careful study, but this is not true of the psychasthenic.

The personality of the neurasthenic is also markedly different from that of the individual who develops psychasthenia. The former has been described above, the latter is essentially a weakling and accomplishes but little in life, being always more or less dependent upon others and unable to reach a decision. Persons suffering with this disorder, like the hysteric, the dementia præcox patient and even the cyclothymic, thus seem to be, from the start, more definitely incapable of adjustment and it is for this reason that these conditions are spoken of as being more constitutional in character and hence less hopeful in prognosis.

Complications and Sequelæ.—The complications and sequelæ which may result from the neurasthenic behavior have already been sufficiently indicated in describing the symptoms. The most serious, as regards prospects of life, are those connected with the limitation of food and the general unhygienic mode of life which may result in actual starvation and exhaustion or the liability to invasion with the diseases of inanition. These need no further discussion here.

Association with Other Diseases.—The development of neurasthenia in association with other diseases has also been sufficiently indicated. But we may here point out briefly certain associations with structural anomalies which have been, and still are, the source of much unnecessary suffering and disability because of the failure to realize the real relation

which they bear to the functional disorder. It is impossible to consider them all as they reach into every sphere of medicine and attention will only be given to some of the more frequent.

The first of these concerns certain so-called malpositions of viscera. Renal, gastric and intestinal ptosis have been greatly emphasized as of frequent occurrence in neurasthenia and have been alleged as the cause of the various symptoms of the disorder by some writers. The absorption of subperitoneal fat which occurs as the result of the prolonged starvation from which many of these patients suffer will necessarily permit exaggerated movement of many of the viscera. Apart from this it is at least doubtful whether visceroptosis is more frequent in neurasthenics than in other persons. There is, however, no question that a neurasthenic who is once informed that he has a moveable kidney or displaced stomach will seize upon this structural finding with avidity and readily adopt it as an explanation for all his troubles. He will willingly submit to exercises, bandages or operations designed to remedy the supposed defect and his actual recovery, when rational therapy is initiated, will inevitably be retarded by reluctance to give up the structural anomaly for the less willingly acceptable functional explanation of the origin of the disability.

In the female, it is only too often that the physician discovers upon examination that there are lacerations or erosions of cervix or perineum, that the uterus does not quite have the so-called normal position, but is tilted forwards or backwards, is antero- or retro-flexed, or there is some degree of prolapse. Any of these may be true and yet give rise to no symptoms at all. But their discovery in the neurasthenic with the assurance which often goes with it, that here is the cause of all symptoms, is very liable to have exactly the same effect upon the patient as is the institution of dietetic measures in the treatment of the gastrointestinal symptoms. The patient becomes confirmed in her fears of a real disease and submits to operations or the wearing of instruments and is thus taught to pay closer attention to her various uncomfortable sensations instead of being assured of their true significance and lack of importance. Probably no one factor is a more potent source of chronic and confirmed invalidism than these mistaken diagnoses in the region of the generative organs.

The plastic operation or the pessary may relieve the patient for a time, as may the elimination of certain foods from the dietary, but the symptoms are not due to malposition of the uterus and consequently relief is only temporary and the failure to cure can result in nothing but increased hopelessness and more settled habit of invalidism.

In neurasthenics, as in other persons, slight deviation of the nasal septum and variations in the turbinates occur which have no real significance, though they can very easily be made the basis of an explanation for his symptoms, readily acceptable to the patient, if the physician gives this trend to his thoughts. Such errors in treatment, made doubtless in all good faith, are extremely frequent and also most unfortunate. The patient is only too liable, as the result of the hope engendered by

the confident assurances usually made by the physician, to be still more confirmed in his disability by the failure to relieve by local applications or operations, which must inevitably occur even if temporary relief is secured.

Some consideration should also be given to the possible association of neurasthenia and tuberculosis. It must be admitted at once that the two conditions are not incompatible. As we have already stated in discussing the etiology, active tuberculosis may, like other infective conditions, even serve as an exciting factor. Again the starvation and unhygienic mode of life to which the neurasthenic is liable may well serve as a factor in the causation of tuberculosis. These associations are, however, certainly rare and hence can have no bearing upon the real nature of neurasthenia.

Clinical Varieties.—Subdivisions of neurasthenia into various clinical types, mainly for convenience in description, have been attempted by many authors. Thus we find such descriptive titles as cerebral, spinal, gastric, cardiac, sexual, etc., neurasthenia. But such efforts at precision in classification accomplish no useful purpose and from what has been said it is obvious that the cerebral neurasthenic of to-day may be a gastric neurasthenic to-morrow. Running through all varieties is the same underlying mechanism and the disorder is essentially one of behavior and not of individual organs.

Treatment.—Neurasthenia being a psychogenic disorder, it is obvious that treatment must be psychic in character. In order to grasp the full meaning of this statement the reader is again referred to the definition of functional disorders given at the beginning of this article. We there found them to be faults in adjustment to conditions of life and hence it follows that recovery means the adoption of modes of reaction which will be satisfactory. The patient needs to be shown wherein his error lies and how to rectify it. This is not a simple task which can be performed in a few minutes by the writing of a prescription or the performance of an operation, but requires painstaking investigation of the actual facts and a whole-souled devotion of time, skill and tact upon the part of the physician without which no permanent results can be hoped for.

In this connection it is worth while to emphasize, as Dubois does especially well, the distinction to be made between what has been called *suggestion* and the very different method known as *persuasion*. To the latter alone really belongs the name of *psychotherapy*, although the former also makes use of psychic mechanisms. But *suggestion* merely attempts, by authoritative reiteration, to introduce a belief into the patient's mind which, if accepted, will counteract the beliefs and fears which constitute the disorder from which he is suffering. *Persuasion*, on the other hand, consists in the actual instruction of the patient in the mechanisms by which he has come to behave as he does, these mechanisms being determined by an actual analysis of the facts of the development of the disorder as given by the patient himself. The former is a short cut, closely akin to trickery; even though performed with the

very best motives, and being founded upon assertion rather than demonstration must of necessity be liable to be swept away.

Suggestions may be made in many ways, consciously or unconsciously, on the part of the physician and with or without hypnotic sleep. The garb in which they are clothed is selected with a view to the impression of the patient. Given with the assurance of the noted specialist, the patient may be convinced by drugs, by electrical contrivances, by surgical procedures, dietetic régimes and what not. The author accidentally met, on one occasion, a neurasthenic who had been relieved of profound gastric disturbances, which had culminated in a strictly milk diet for two years, by following the advice, skillfully given with assurance, that coffee drinking would cure him. He rapidly improved and was soon on a full diet because, so he believed, he drank two cups of coffee with every meal. The outcome of the case the writer does not know, but one can readily appreciate the shifting nature of the sands on which the improvement was built.

That temporary and even prolonged improvements in symptoms can be produced in this manner there is no doubt. We have but to observe the results undoubtedly secured in cases treated by the various medical and religious cults and fakers. But such cases, whether treated by regular or irregular practitioners, cannot be regarded as cured even though they may be relieved for a longer or shorter time.

It may save time and trouble to give tonics, bromids, static head breezes or other suggestions, hypnotic or otherwise, but it cannot be too strongly asserted that medicaments and suggestion have no place in the treatment of neurasthenia, whatever be their status in regard to other functional disorders. There is only one method of rational therapy and that, time-consuming and troublous though it may be, is **persuasion or education**. This is not intended to imply that drugs, diets and physical remedies may not properly be employed in combating the secondary conditions of anemia, starvation and exhaustion which may result from the neurasthenic mode of life, but these are directed towards the relief of complications and not towards the cure of the neurasthenic condition itself.

The first stage in the treatment begins with the examination of the patient which must be especially thorough and complete, not only to avoid errors in diagnosis, but also with the object of satisfying the patient that the physician is fully informed of all the facts, knows his business, and is therefore a competent and conscientious teacher. The establishment of such relations of confidence and esteem is absolutely essential to obtain a reliable history of the origin and evolution of the disorder, without which the physician can do nothing but treat symptoms.

In the absence of physical findings, which will be determined by the ordinary routine methods of examination, the main study must be directed to the patient's account of his illness. This invariably takes considerable time and may require several interviews, facts which have been omitted from the account at first, with or without intent, often

being recounted in later interviews when confidence in the physician's genuine interest has been established. This probing into the history should be conducted openly and frankly, without special mystic arrangement of the conditions by semi-darkness, reclining posture or other hypnoidal measures, for it is, the writer believes, most important that the patient be fully conscious and that he reach his own conclusions. Suggestive influences, such as those mentioned, may bring more rapid results but they are liable to be less fully accepted by the patient and therefore less effective.

During this first stage it is well to permit the patient to tell the story in his own way with but little comment and that directed merely to encouraging a complete unburdening. When the patient seems to have related everything, questions concerning the presence or absence of manifestations in other organs, prompted by the physician's experience and knowledge of what is probable, will often serve to bring to light earlier experiences which had been forgotten. The greatest care should be exercised to convince the patient throughout that the physician realizes the importance of the various symptoms and to avoid any suggestion that he regards them as imaginary or unreal. At the same time, here, as in the physical examination, it is important that there be no suggestion of any desire to hide anything from the patient.

The time spent in this process, while it may seem tedious, is not lost and may, as the direct result of confiding his troubles to some one else, bring about a very definite improvement. It is also important to secure the whole story if one is to avoid the possibility of having the patient take refuge from the true but unpalatable explanation of his reactions, which represents the purpose of this study, by bringing up certain symptoms, not previously described, which may seem to contradict the views advanced.

The next step in the therapeutic procedure consists in reconstructing, from the facts given, the mode of development and real significance of the various subjective manifestations. These we have already learned to recognize as exaggerations of emotional adjustment and the patient must be convinced, by the facts of the evolution as he himself has given them, of their relation to the conflict which has been the starting point. This means that he must come to acknowledge that he has made use of the sensations belonging to the unpleasant emotion of the conflict to provide a means of escaping from the need for continuing the fight. Such a realization is not going to be achieved willingly at first and he may use much ingenuity in attempting to find some other explanation which would avoid the need for again taking up the burdens of responsibility.

The attitude of the physician towards this resistance must be one of encouragement and sympathy but any effort at suggestion should be avoided. The patient should be made to understand that the disappearance of the various symptoms rests with himself and his willingness to relinquish them. It is not a question of his forgetting the uncom-

fortable feelings, pains and disabilities but of his really facing them and grasping their nature.

Such, in general, is the outline of the fundamental basis of treatment of the neurasthenic state but it is necessary to be somewhat more specific in regard to the actual **management** of certain aspects of the disorder. First among these may be considered the question of the removal of the patient from the conditions under which the neurosis has developed. Experience in the war demonstrated beyond question that the best results were secured by treatment as near the firing line as possible. This should be applied to the conditions of peace. But it is necessary also to take into account the fact that nothing is more conducive to the maintenance of the disability than the solicitous devotion and sympathy of relatives or others, convinced of the existence of bodily illness and structural incapacity, for this attitude must inevitably tend to confirm for the patient his own fears and wishes.

For these reasons alone it is most often absolutely essential to remove the patient from his home and to place him under conditions in which he will meet with nothing that will not tend towards squarely placing the responsibility for the symptoms where it belongs. Occasionally it may be possible, in less advanced cases, to avoid this removal, but even then it will often be found that the relatives require more care in the way of education than does the patient himself. Unless there is good reason to believe that this can be satisfactorily accomplished, removal to a hospital should be a *sine qua non* of treatment.

In the hospital the **complete isolation** of the patient, especially from friends and associates, first recommended by Weir Mitchell, is often advisable for two reasons. First, to permit the full accomplishment of the purpose stated in the last paragraph and, second, to provide temporarily for complete relaxation and to convince the patient that the illness is being taken seriously. It also affords an opportunity for thorough self-analysis. The character and training of the nurses who are to care for the patient during this period are obviously of the utmost importance. Some knowledge of the nature of the disorder is absolutely necessary and must be combined with a quiet assurance and sympathetic tact which must be entirely free from all sentimentalism.

The actual duration of the period of isolation, which should be accompanied by complete **rest in bed**, will vary with the severity of the disorder and the improvement shown by the patient in his attitude towards the symptoms. Usually from two to six weeks is sufficient to render the assumption of more active life, with a gradual return to full responsibility, permissible. During the period of rest in bed, circulation and nutrition should be maintained by the use of general **massage** for twenty or thirty minutes daily, the vigor and duration of the rubbing being gradually increased from day to day, care being taken not to carry the stimulation to the point of fatigue. As soon as possible the interest of the patient should be attracted to more active participation by the application of **occupational therapy**. This may be carried out for short periods at first, gradually increased within the limits of fatigue.

The earlier types of occupation selected should be distinctly sedative and more or less monotonous, but as improvement occurs they may be made more varied and such as call for closer attention upon the part of the patient in their execution. In these later measures consideration may be given to the interests peculiar to the patient with the object of providing him with a hobby and means of recreation which may be continued and be of inestimable value when he returns to a normal life.

The period of isolation, during which the psychotherapeutic conversations and explanations are carried out, is also one in which attention should be given to the **education of friends and relatives in the attitude towards the patient** which must be adopted when he returns to home and work. It is very important that they realize that the patient is coming back fully restored and that his ability to undertake the responsibilities in life must not be questioned in any way. He is not an invalid and his chances of successful readjustment will be seriously jeopardized by any unwise solicitation. They must learn that the capability of the patient to meet the difficulties in life, inherent in his particular business or profession, have received the most careful consideration by both patient and physician and that the latter is absolutely sure of his ground, whether a change in the previous mode of life or occupation of the patient has been decided upon or not.

Following this period of isolation, the **sphere of activity** of the patient may be **gradually extended**, relations reestablished with the outside world and more vigorous exercise encouraged. These should always be kept short of the point of causing fatigue. At this time, too, it is well to begin the establishment of hygienic measures which are to be continued in the future life of the patient, such as systematic exercises, shower baths, etc. In this, as in all other measures, it is important that the patient realize that he is only learning measures of health and is not receiving remedies for any disease.

With regard to the treatment of individual symptoms, too great stress cannot be laid upon the necessity for avoiding any measure which could be accepted by the patient as the cause of his improvement or relief, for such can only serve to confirm a belief that some structural disease was present. Thus, for instance, *hypnotics should not be given to relieve insomnia*. If the patient has been in the habit of taking them he may be allowed to continue them but must be taught that even though they perhaps make him sleep they do not and will not really help his condition. Successful psychotherapy will convince him that his failure to sleep is due, not to any inability to do so, but to his adoption of insomnia as a relief from something more intolerable, unpleasant and fear-some though it is. He can sleep if he will permit himself to do so. Even *hydrotherapeutic measures should not be prescribed as hypnotics* though they may well be used upon the understanding that, like isolation and rest in bed, they are part of the routine measures designed to relieve the secondary effects of his neurasthenic mode of life.

Severe gastric disturbance must also be met in the same way. *Drugs and digestive enzymes must not be used*. The patient must learn, as he

speedily will with proper explanation and encouragement, that he can digest his food if he will. Such patients generally are greatly undernourished and suffer more or less severely from the consequences of partial starvation. The remedy is **forced feeding**, gradually but rapidly instituted and insisted upon with quiet assurance. Like the avoidance of fatigue insisted upon above, it is important that the addition of articles of food to the dietary be made with due consideration for the condition to which the stomach has been reduced by habits, more or less prolonged, of insufficient food. But it is rarely that full mixed diet with, in addition, three quarts of milk daily cannot be reached in two or three weeks. Constipation must be met by the establishment of regular habits rather than the administration of purgatives.

The *treatment of coexisting diseases or defects* must depend upon the urgency of the complicating factor. If the principles of psychotherapy laid down above have been fully grasped it will be obvious that the absence of any real relation between the defect or disease and the neurasthenic condition must be fully established to the satisfaction of the patient and the author cannot too strongly deprecate the performance of plastic operations or the application of instruments simply because they might do some good. The patient is only too anxious to find some structural anomaly to explain his condition and anything which will tend to exaggerate the importance of some trivial displacement or deformity will surely result in the postponement of real recovery.

Prognosis.—The prognosis as to complete recovery and restoration to full functional activity is essentially good, provided proper treatment be adopted. Occasionally the disorder may disappear spontaneously when the conditions which brought about the conflict cease to operate sufficiently early. Many illustrations of this fact were afforded by the rapid recovery which followed the signing of the armistice in the recent war. But, as a rule under peace time conditions, the probability of such spontaneous recovery is not great. The duration of the disorder is therefore extremely variable and it may be said in general that the longer the disorder is continued, and hence the more deeply established the habit of reacting in this way, the less favorable the outlook as regards recovery.

Neurasthenia does not tend to develop into insanity, although the need for care in diagnosis here must be especially emphasized, even though the disorder continue to exist for many years. Furthermore, true neurasthenics do not commit suicide in spite of the fact that this name is sometimes applied to disorders which have led up to this unfortunate ending. Such errors are sometimes the result of fault in diagnosis and at others the name is used in order to avoid the greater stigma which popularly attaches, very unjustly, to insanity in any form.

Occasionally death may result from the severe exhaustion which results from starvation and unhygienic modes of life, but such eventuality must be regarded as an accident and is certainly very rare.

Pathology.—From the tentative definition and the description of the disorder already given, it will at once be obvious that neurasthenia

presents no morbid anatomy. Under the heading of pathology it therefore only remains to discuss further the pathogenesis and nature of the disorder and to attempt a more exact definition.

The essential nature of neurasthenia has been the subject of much controversy and, while there are many modifications as to detail, the theories which have been developed may be considered under three main headings. These we may label (1) toxic, (2) defect in development, and (3) psychic.

(1) The close resemblance which exists, superficially, between the irritable weakness resulting from severe bodily intoxications and the fatigability and hypersensitiveness of the neurasthenic state has naturally led to the assumption of the presence of a toxic state as the explanation for the development of neurasthenia. This theory has, in addition, the advantage of providing a structural basis for the disorder and thus of satisfying the materialism with which most physicians are imbued. It is rendered plausible by the occasional development of neurasthenia following some infectious disease. In the absence of such demonstrable intoxication it is always possible to fall back upon some hypothetical autotoxin, fatigue product, endocrine gland disturbance or what not.

But it must be admitted that no evidence has yet been produced, beyond the existence of the symptoms for which we are seeking an explanation, that any toxin is present in even the majority of cases. The disturbances in organ function which are found, are throughout quantitative rather than qualitative and it is at least difficult to conceive of a prolonged, as many of the cases are, intoxication of tissue without the occurrence of definitely demonstrable structural alterations. Since, in addition, the facts are fully capable of explanation without any such hypothecation, it seems logical to conclude that the burden of proof must rest upon the exponents of the theory and that, until proof of the existence of a toxin is forthcoming, this hypothesis may be ignored.

In drawing this conclusion it is not intended to deny that autotoxins may arise as a consequence of the neurasthenic mode of life, the possibility of which, as of secondary infection with tuberculosis, was pointed out, for instance, in speaking of the acidosis which may arise from the partial starvation. But here, there is no question of the toxin as the cause of the disorder.

(2) Another theory, which has been very widely accepted and which again endeavors to establish a structural basis for the origin of neurasthenia, would include it with those conditions which are based primarily upon faulty construction. Such a view does not conflict with the toxic theory nor with the concept of a truly psychic origin to be described in a moment.

That all persons are not equally endowed with the ability to withstand stress, be this toxic or a prolonged demand upon the functional vigor of the organism, needs no argument. Experience during the world war seems strongly to suggest that even most carefully selected and, judging from their past record, vigorous and healthy individuals may

develop neurasthenia, provided the stress be sufficiently great. It seems therefore hardly a sufficient explanation to allege that the individual who succumbs was of inferior constitution.

The earlier history of many persons suffering with a neurasthenic state will reveal no special reason for anticipating a breakdown, a fact which of itself seems to suggest that there is probably some essential difference between this condition and other types of functional disorder in which the constitutional factor is far more constant, such as the hysteric, the psychasthenic and the paranoic types of reaction.

While therefore we recognize that poorly constructed persons break down with neurasthenia more readily than do those of more sturdy build, yet this is of itself not a sufficient explanation, and it becomes necessary to find some other factor which can be sought only in the stress itself.

(3) The psychogenic theory requires no hypothecation of structural alteration beyond that which accompanies emotion, and is physiological in character, even though structural damage may result secondarily. According to this view the individual finds himself placed under conditions to which he is unable to discover a satisfactory solution. By this is meant that, under the restrictions placed by social requirements (as he understands them), the situation is more or less intolerable to his instinctive cravings. This implies worry and apprehension with their accompanying bodily disturbances which normally give rise to conscious perception.

Bodily illness is a good reason for retiring from active participation in the work of the world, not only permissible by social usage, but acceptable to the most conscientious individual. Looking for a way out of his difficulties, the man is liable to grasp eagerly at any straw which offers such a logical excuse for "quitting," and there is needed but the suggestion that the uncomfortable feelings which belong to the state of emotional upset are indications of disease. It is true that the illness means much suffering and discomfort but it brings relief from something more intolerable. The selection is not made with conscious realization of the true meaning of the step which is being taken, for uncomfortable sensations and pains are already actually present as part of the emotion though they will become exaggerated by the attention given to, and the interpretation placed upon, them.

How the suggestion, which precipitates the breakdown, is given will vary greatly in different cases. But, as an illustration, one may mention the solicitous devotion of relatives and friends concerning the worried look, the loss of sleep and lack of appetite, with concomitant loss of flesh which come to every one in times of emotional stress, if long continued. The sufferer is advised to take care of himself, not to work so hard, to take a rest or to see a doctor. This advice may be reiterated many times and be emphasized by special attentions of all kinds. Such solicitation may be greeted at first with irritability but is none the less acceptable and may soon come to be demanded. Chance remarks of friends, the reading of some advertisement or a visit, more or less shame-

facedly, to the doctor may tend to reinforce the suggestion which finally takes abiding root in the patient's mind. He begins to observe his sensations more closely and to study the disturbed functions of his body organs with the inevitable consequence that they grow steadily worse. Should there develop at such a time some real, even if temporary, illness, the relief experienced in consequence of the retirement from irksome responsibilities, thus compelled, may obviously serve as a most potent suggestion.

But, it may be asked, if this mode of development be accepted as a satisfactory explanation of the onset, why is it that the individual does not recover when he is removed from the conditions which gave rise to the conflict? The answer to this question is that not unfrequently he does recover. When he does not, various factors may be concerned in the fixation of the reaction. First, recovery might necessitate a return to the old difficulties. Secondly, the dominance of his surroundings, the solicitous care and the general freedom from responsibility which accrue from the disability may serve as a very potent argument in favor of not getting well. These mechanisms were admirably illustrated in the earlier cases of war neuroses among the British and French in the recent war.

A third possibility, one which has an important bearing upon prognosis and treatment, is that the reaction, if continued because it secures desirable results for any length of time, may, like other ways of doing things, become a habit. The formation of habits of reaction is one of the special functions of the nervous system and means nothing more than the fact that paths of association frequently used become progressively more easy of access and thus permit great saving of time and energy by avoiding the necessity, in future, for selecting the response to any given stimulus. Habits once established are proverbially difficult to break in proportion to the frequency of their employment. Habitual neurasthenic reactions are no exception to this generalization and such cases are very resistive to treatment. They may, indeed, never be entirely eradicated.

The foregoing discussion will now permit us to consider again the definition of *neurasthenia* upon which must be based the question of differential diagnosis. The essence of the disorder, as outlined, consists in the adoption of a mode of reaction to conditions of conflict between social restriction and instinctive needs by accepting the awareness of those disturbances of organ function, which are part of the emotion aroused by the conflict, as evidence of bodily sickness and thus as a reason for retiring from the conflict.

The first part of this definition, as already explained, is covered by the phrase functional nervous disorder. The particular form of the disorder is characterized by the use which is made of the unusual sensations of quite normal body activity.

The definition may then be phrased as follows: *Neurasthenia* is a functional nervous disorder characterized by the acceptance by the patient of the subjective equivalents of disturbances in organ function

inherent in emotion as evidence of disabling disease and thus as a means of escape from some intolerable situation.

PSYCHASTHENIA

Synonyms.—Constitutional neurasthenia, Zwangsneurose, Aboulia, Folie de doute, Krishaber's disease, Phrenasthenia, etc.

Definition.—The word psychasthenia means absence of mental strength and was first used by Janet to designate a group of functional nervous disorders characterized by doubt and hesitation with obsessions, fears and impulsive acts.

Etiology.—**PREDISPOSING CAUSES.**—*Sex.*—Psychasthenia is certainly much more frequent in women than men, the proportion, according to Janet, being about three to one.

Age.—The disorder is essentially one arising in early life, the great majority of cases being fairly well established before the age of 20. But it is not unusual to meet with cases in which the symptoms become obvious only in the later years of life. Even in such instances, however, the life history will almost invariably reveal evidences of over-scrupulousness and rumination as more or less characteristic traits of the individual's make-up. Age seems therefore only to be of importance because of the varying degrees of difficulty in social adjustment which come with development and advancing years. The period of puberty with its enormous demands for adjustment because of the wealth of new, inherent cravings, peculiar to sex development, soon followed by the added responsibilities belonging to the adult and the necessity for emancipation from home and parental control, must act as an extremely efficient test of the capacity for social adjustment possessed, either as the result of inheritance or of training, by any individual. It is therefore not surprising that it is at this stage of development, especially, that poorly equipped persons show in unmistakable form their inability adequately to meet the demands made upon them. Such deficiency in equipment is often spoken of as constitutional, although it must not be forgotten that the quality of the education received is a factor to be considered as well as the quality of the tissues which the individual has inherited. The period of adolescence is the one in which all such disorders, including hysteria, dementia præcox and psychasthenia, tend especially to develop, not because they are inherent to the chronological age, but because it is at this time the first real adjustments to life and individualization must be accomplished.

Heredity.—That heredity is a most potent factor in the development of psychasthenia is asserted by all writers, insanity, psychasthenia, alcoholism or neuroses being present in the family history of at least 90 per cent. of the cases. According to Pitres and Régis this hereditary taint was of similar kind to that of the patient in 39 per cent., while Janet found such relationship in 28 per cent. Tuberculosis, syphilis and chronic arthritic diseases have been quite frequent in the parents.

In some instances there seems to be a relation between the development of psychasthenia and advanced age of the parents at the time of conception. According to Soukhanoff transmission is more frequent from mother to son and from father to daughter than to the offspring of the same sex as the affected parent.

In this connection it is worthy of note that many of the earlier authors laid great stress upon the frequency of the so-called physical stigmata of degeneracy. That some psychasthenic individuals do show an unusually large number of such abnormalities is certainly true; but many of them, probably the majority, are well formed and show no greater proportion of stigmata than do normal persons. This conclusion was reached by Pitres and Régis after an exhaustive study of this feature of the subject and was concurred in by Janet.

The influence of heredity in causing such a disorder, especially where the inheritance is like in kind, is rendered more difficult of determination by the fact that the children of a psychasthenic person are also very liable to be trained to indecision and procrastination as the result of association and example. Yet, even so, the existence of some special defect seems to be suggested by the fact that children, apparently quite healthy, may grow up in the same family with a psychasthenic child.

Personality.—It is becoming customary to ascribe to the personality of the individual, that is to say the manner of reacting to situations which he develops, and which are therefore characteristic of him as an individual, an important rôle in the predisposition to functional disorder. But, in reality, if such disorder is truly constitutional, as defined above, it is obvious that the disorder itself is nothing but an extreme expression of the mode of meeting difficult situations or, in other words, the personality of the patient. To describe the personality is therefore to outline the kinds of reaction which, if carried to their logical conclusion, become the "symptoms" of the developed disorder. Nevertheless, if prevention of such developments by appropriate treatment is to be attempted, it is essential to outline clearly the slighter and earlier characteristics of the behavior which is liable to evolve in this manner. With this understanding of the true relationship between personality and the constitutional types of functional nervous and psychic disorder, we may devote some space to describing the personality of the individual liable, under sufficient stress, to become psychasthenic.

The usual history which one obtains describes these persons as of especially sweet and gentle disposition. As children they were always well behaved, docile and never caused a moment's trouble. Such a child is not normal; mischief and the frank expression of feelings and emotions are natural attributes of childhood. The real reasons which lie back of the failure to cause trouble are the identical factors which make up the psychasthenic reaction. Such persons are extremely timid and self-conscious, find the utmost difficulty in reaching decisions, and will suffer great personal inconvenience rather than struggle to alter the conditions. Change and action are abhorrent with the consequence that there is a marked tendency to extreme precision and exactitude. Everything must

be kept exactly in the same place and all things and acts must be arranged in some definite and more or less unchanging order.

The need to reach any decision is a matter of great difficulty and these persons hesitate, ask advice as to the value of which, when given; they are unable to decide; they adopt any course which holds out a hope of postponing action. Their acts are hesitant and liable to be incomplete and they tend to worry whether it would not have been better to have done something else. In consequence they are often clumsy and maladroit. They show marked bashfulness and difficulty in appearing in public or meeting strangers, will stay silently by themselves and avoid so far as possible all occasion for active interference. They are easily led and always subordinate.

Their interests show the same tendency to keep away from contact with situations calling for definite decisions and they delight in the vague and the abstract. As Janet says they adore poetry, philosophy and religion; "they live in the future and above all in the past rather than the present." Correspondingly they are incapable of deep emotion, the general trend of mood being a quiet melancholy with marked timidity. They react to demands for decision by becoming restless, agitated and fearful. They form no real friendships though they may be tolerated by many acquaintances.

Intelligence and clear grasp of the situations in which they find themselves are usually above the average. They do well in school studies, especially the more philosophic subjects, and since they prefer to read and study, rather than to take part in active games and amusements, it is often alleged that they work too hard and that this is the cause for the breakdown which appears later.

The principal characteristics are thus mainly negative and the dominant feature in the whole picture is one of indecision and doubt (aboulia) with avoidance of positive action.

EXCITING CAUSES.—In very many cases it is difficult to determine any one factor which seems to have acted as the precipitating cause. Every demand for the assumption of fresh responsibility brings with it an increasing liability that the indecision will become so manifest that a continuation of active participation in life is much interfered with or is no longer possible.

The actual breakdown, that is to say the moment when friends or relatives first realize that there is something definitely wrong, usually comes at some moment when the patient is finally obliged to make some more or less important decision, be this to select some means of earning a livelihood, to leave the parental roof or to get married. The patient may have allowed himself to drift into an engagement because this was easier than to say "no" and required at the time no definite action, but the moment for active decision and participation must inevitably follow sooner or later.

Occasionally the precipitating factor appears to be an emotional shock, a horrible accident witnessed, a disappointment in love, the loss of a husband, etc. In such cases the disorder may appear to develop

almost suddenly but even then it is rare that there has not been more or less well marked evidence of a psychasthenic personality throughout earlier life.

Symptomatology.—**CLINICAL HISTORY.**—From what has already been said it will be appreciated that the onset is, most usually, extremely insidious and it is very difficult, if not impossible, to give a date when the malady can be said to have begun. In rare cases it is more abrupt and instances of sudden onset at the moment of some emotional shock have been recorded. The symptoms are so very largely subjective that they may be in existence for a long time without being made known to others. The indecision and hesitation which are so characteristic of the patient also tend to prevent him from avowing the existence of symptoms, distressing as they may be, until they can no longer be concealed because of the effect they produce upon conduct. These patients, as a rule highly intelligent and discerning, also most often have full insight into the subjective nature and more or less absurdity of their difficulties with the consequence that they are ashamed of them and fear to arouse ridicule and public attention. This also tends to keep back the final avowal and open display of the fears and doubts with which they are tortured.

Once recognized there is a tendency for the difficulties and rumination to become progressively worse in the sense that they interfere more and more with the ability of the patient to continue in his field of work. Exacerbations, amounting to veritable crises, upon the occasion of unusual demands and remissions when events go more smoothly and evenly are very frequent as in other forms of functional disorder. There is, however, a marked tendency for the preservation of the symptoms in the same form even though the disorder continue for many years. The manner of spread is rather in the direction of the development of wider associations so that the particular difficulties in thought, feeling and action which make up the disorder are brought to consciousness by more and more of the events and situations of life.

Subjective Symptoms.—The manifestations of psychasthenia are essentially mental and hence we shall look for them especially in the realms of thought, feeling and action. Janet has given a most accurate and detailed account of these phenomena, based upon the study of a large number of cases, upon which it is impossible to improve. We shall therefore follow, more or less closely, the outline which he has given without, however, detailing the elaborate classification he has employed. It must be borne in mind, in reading this description, that, while the disturbance in some one of the three spheres of mental operation may seem to be most prominent in any given case, there is no real separation between them and all are inevitably more or less involved.

Certain features present in all manifestations stand out with especial prominence. In every instance it concerns an obsession, that is to say the obtrusion into consciousness of thoughts, feelings or tendencies to action which appear to be more or less foreign to the general stream of thought and action and strange to the setting in which they arise.

They are felt and described by the patient as uninvited and unwelcome guests, injected, as it were, from the outside, unpleasant and yet absolutely compelling in their demands for attention. They seem to be unescapable and unending, a continual mulling over and over which constitutes a veritable mental rumination.

Obsessive ideas (*les obsessions, Zwangsvorstellungen*) are grouped by Janet into five classes according as they seem to relate to topics of (1) sacrilege, (2) crime, (3) personal inadequacy, (4) bodily deformity, (5) hypochondria. The distinction between these classes is not, by any means, always clear and the same patient may have obsessions belonging in several or all groups. Illustrations of the character of these ideas from the author's cases may be cited as follows: (1) the recurrence of ideas of the male genitalia whenever anything relative to religious observance was brought into consciousness, going to confession, talking to a priest, a picture of the Madonna and Child, a crucifix or the mere thought of them aroused in any way. (2) The idea of her husband murdered by herself whenever she saw a knife or any sharp implement or even any object resembling in shape such a weapon. (3) The thought "I am almost an idiot," "I have no comprehension, I am useless and unfit to live." In this same category Janet also includes two very striking types of obsession which have been the subject of much discussion. The first of these concerns ideas of loss of personal identity (*dépersonnalisation*) in which the individual seems to be not himself; he acts, as it were, without volition and feels as if he were merely an onlooker. The second contains the idea that everything which happens is familiar and must have occurred before (*déjà vu*). (4) Thoughts of grotesqueness in appearance as the result of excessive hair on the face, being too fat, blushing so readily, of great clumsiness in action. (5) Ideas of sudden death, of tuberculosis, heart disease, cancer, of being infected with syphilis and hence a menace to others, etc.

All these ideas are accompanied by depression and apprehension and many by a strong impulse to action in accord with the obsession which, however, never follows if it be at all dangerous. The patient outwardly may give little or no sign of the distress he is experiencing or may show a restless agitation which at times may be almost frenzied.

Closely akin to these obsessive ideas are a group of doubts and hesitations (*les agitations mentales, les manies mentales, rumination mentale*) which may be purely thoughts or be accompanied by ruminative acts. Like the obsessive ideas and other manifestations, their absurdity is acknowledged by the patient, but he feels obliged to continue them monotonously often for hours at a time. Thus a patient of the writer's spent hours and days continually dressing and undressing. Whenever she sat down at a table she was obliged to get up and sit down again many times. These acts had to be repeated an odd number of times and she kept careful count of them. Another feels compelled to repeat over and over various conversations she has had during the day, or as she puts it, "What I see, I see again, what I hear, I rave." Things which have been done on a certain day in the week must be done again on

the same day of the week or month, else "something terrible" will happen. The fear of dirt may lead to innumerable repetitions of sweeping or washing. The flagstones in the pavement must be counted and every fifth one avoided, etc.

Sometimes the ruminations are related to visceral activities, especially the stomach, and repeated vomiting or efforts to vomit, because of the fear that the food was indigestible, will cause flatulence, etc., are very frequent. The bladder functions may be similarly interfered with and masturbation may be practiced incessantly.

Obsessions in the realm of feeling (*les agitations forcées émotionnelles*) may be either general and diffuse or more specific (*systematisées*). In the former case there is a general state of apprehensive anxiety, a feeling of impending danger or disaster, always more or less present but varying in intensity from time to time. The latter are usually termed phobias and considerable ingenuity has been exercised by various authors in devising special titles to cover particular varieties of fear. Janet groups them into four categories: (1) fears of the patient's body which include many of the fears belonging to the obsessive ideas referred to above; (2) fears of outside objects: dangerous articles, contamination (mysophobia), of individuals, animals, etc.; (3) fears of physical or social situations: lightning (astraphobia), places (topophobia, agoraphobia, claustrophobia), fears of blushing (erythrophobia), of society (anthropophobia), etc.; (4) fears of particular ideas: religious or moral thoughts, fears of fears (phobophobia), fears of death and of disease (pathophobia, syphilophobia, etc.).¹

Obsessions in the sphere of action (*les agitations forcées motrices, Zwangsvorgänge*) represent activities which the patient feels obliged to carry out in spite of himself. Like the emotional disturbances they may be either specific in the form of ties or more general and diffuse when they constitute a general agitation.

Tics are sudden and abrupt movements which may involve the muscles of the face, eyelids, mouth, tongue, pharynx, trunk or limbs. They occur spontaneously and without relation to the situation at the moment. They originate in a definite act, responsive to a more or less adequate demand for reaction but, for some reason, to be discussed later, are repeated without cause and gradually lose their completeness and become simplified, by the omission of certain stages of the act, to muscular contractions which in appearance are quite purposeless and very often odd and grotesque. Thus, for example, the movement of clearing the throat, due at first to inflammation and dryness of the throat, if for any reason it become an obsession after the throat has recovered, may degenerate into a mere movement of the palate and expiratory muscles resulting in a clicking noise. Tics therefore accomplish nothing either good or ill. Many of them represent only the remnants of the ruminations which we have already considered.

In addition to the tics resulting in movement, Meige and Feindel point out that there are tics in which the muscular contractions are tonic and result in the adoption of some attitude of body. Thus, while walk-

ing, the patient may suddenly stop with one foot raised and body poised for a brief period every few steps.

In the diffuse type of agitation, the patient gradually works himself up into a state of restless activity, repeating monotonously certain movements such as walking, weeping, striking himself with his hands, uttering inarticulate cries like some animal, etc. Sometimes these crises of agitation show almost a choreiform character of muscular contraction and such have been described as "electric chorea."

Emphasis has been laid upon the fact that the patient does not actually lose touch with reality and is aware of the subjective nature and absurdity of the obsession. Occasionally, however, it almost seems as if there may be, for a time, a real loss of orientation and the obsessive ideas thus become hallucinations. There has been considerable discussion upon this point and the general consensus of opinion is that there is never any true objectification of the ideas such as occurs in hallucinosis. A patient of the author's, who experienced the impulse to utter obscene words to persons for whom she had the deepest respect, expressed the thought that the devil was controlling her. This she usually admitted to be merely a thought, but in some crises of agitation she asserted emphatically that the devil was standing behind her and that she could hear his voice. Even then she did not convince one that she meant he was objectively present.

Similarly, in regard to the impulses to action belonging to obsessive ideas, assertions have been made that the acts are never carried out. Entirely harmless parts of them may be performed but, except by accident, suicide, homicide, theft, etc., are never consummated. The patient referred to above was heard occasionally to say "I love you" or to curse or use obscene language, *sotto voce*, but only under conditions in which this would not be observed by any one but the physician. None of her friends or associates ever heard any such utterance. Janet also describes an interesting example of a patient who had an impulse to steal silver, etc., when out to dinner, and who was usually accompanied on such occasions by a valet whose duty it was to return these articles. When this attendant was not present he never stole, although the impulse to do so was present.

Another feature which especially needs emphasis is the satisfaction which the patient apparently secures from the performance of the ruminative acts and obsessions. Tics, for instance, can often be controlled by voluntary effort, if sufficient demand is made, but will be followed by what Patrick has called a veritable orgy of tic movements.

This seems also to hold true for the other types, including even the fears. The patient is distressed not so much by the rumination itself as by the interference with other activities and the opinions of other people which result. Under home or habitual surroundings the manifestations are therefore very much more marked than when in the presence of strangers and in unusual situations.

Since depressive emotions and inability to decide play such a large part in the picture it is not surprising that at times there may be added

man, of the symptoms already described under neurasthenia. The mode of origin, however, is essentially different. In the psychasthenic they arise from the obsessions and are not the direct outcome of the state of emotion caused by an intolerable situation.

Sexual Disturbances.—The frequent interference with sex functions has been noted by many authors and deserves special mention for the reason that this association has led to the effort upon the part of Freud and his followers to establish that irregularities in sex life are the source of the condition.

As might be expected from a study of the essential features of doubt and hesitation, of failure to reach conclusions and generally to procrastinate, the sexual life tends to show difficulties and ruminations absolutely in keeping with the other manifestations. Fears as to potency, the consequences of early masturbation, or of bodily deformity or inadequacy, obsessions relating to chance, childhood or later experiences in connection with sexual activity, doubts as to the consequences and their advisability; phobias concerning the possibility of previous venereal infection, etc., all must have a profound effect upon the sexual life. Impotence, incomplete erection, delayed or premature ejaculation, masturbation and even various forms of perversion all occur but should be regarded as symptoms rather than causes.

PHYSICAL FINDINGS.—The psychasthenic individual presents no evidences of organ disease. Physical findings therefore are not part of the picture. As a secondary result of some of the manifestations emaciation and semi-starvation, due to fears of the food, ideas of poison, ruminative vomiting, etc., may result. Calluses, loss of hair, etc., may be developed as the result of obsessive movements and the patient may become actually exhausted by the prolonged repetition of compulsive acts.

The nervous system is characteristically free from evidence of disease. Anesthesia does not occur. The tendon jerks may be very brisk, but there is no clonus. Gastric, intestinal, renal and cardiac functions are unaltered except in so far as they are part of the emotional state.

Diagnosis.—The diagnosis of psychasthenia rests upon the existence of a mental disturbance characterized by the features of doubt, hesitation, obsession and rumination without loss of a full appreciation of the subjective nature of the disturbance.

The chief conditions from which this disorder must be distinguished are, hence, the insanities. The manifestations throughout are so definitely mental that there can rarely be any difficulty in differentiating it from organic nervous disease. A word of warning is necessary in this respect, however, for psychasthenic personalities are subject to diseases just as are more adjustable types and the manifestations of psychasthenia may well become obvious, be avowed or be exaggerated under the stress of illness.

The differentiation from other psychoneuroses is usually not difficult. The symptoms of hysteria are essentially disturbances in body function; the hysteric and the psychasthenic personalities have already been de-

scribed and show a striking divergence. The psychasthenic is also absorbed in his disorder, while the hysteric tends to forget and disregard it. A comparison with neurasthenia has already been made.

Among the insanities especial reference must be made to agitated depressions, dementia præcox and some forms of paranoic reaction. In agitated depression, especially that appearing in the climacteric period but also in younger individuals, there is marked anxiety with a monotonous repetition of painful thoughts, self-accusations and ideas of negation. But, here, one is impressed with the finality and completeness of the conclusions, the tendency to decisive action (e.g., suicide) and the dominance of the depression. The patient is convinced of the truth of his beliefs and does not realize their inappropriateness under the circumstances nor does he even grasp the real situation.

In dementia præcox and some paranoic conditions we also meet with strange and odd thoughts (autochthonous ideas), impulses to action which seem to come from without the patient's own self (pseudospontaneous acts), oddities and mannerisms, ties and persistent mulling over some circle of ideas which may bear a superficial resemblance to those of psychasthenia. But here again the real test lies in the actual disorientation which is present in the former. There is a tendency to real objectification of the thoughts and acts. In the dementia præcox individual there is rarely lacking some evidence of real diminution of interest in the world and associations which sooner or later tends to involve even the dissociations themselves. The paranoic individual has a strong tendency to decisive action which is quite foreign to the psychasthenic.

The importance of making these distinctions cannot be overemphasized, as failure to do so may result in disastrous consequences. Suicide, homicide and self-mutilation may occur in the insanities, but are unknown, except by accident, among psychasthenics.

Complications and Sequelæ.—The possibility of starvation and exhaustion have already been sufficiently indicated. They are not by any means frequent. The possibility of psychasthenia developing into an insanity has been given serious consideration by some authors but, if this ever occurs, apart from the concomitance of organic nervous disease such as syphilis, it is certainly extremely rare.

Treatment.—**PROPHYLAXIS.**—The prophylaxis of psychasthenia may be considered under two heads: (1) the prevention of the birth of persons of this type and (2) the education and protection of children presenting evidences of a liability to such development. The problem of eugenics as applied to man is one which has many bearings and is so complex that it is impossible to discuss it here. While it is relatively simple to decide upon the particular qualities desirable in cattle, horses or other animals and to establish standards in breeding to secure the desired results, it is quite another matter for man, and an absolute standard and general equality, assumed in our constitution, is probably not even seriously desired. Some day it may be possible to establish, with reasonable assurance, that certain traits of behavior should prohibit procreation and

to enforce their observance. For the present we can do little but recommend certain obvious precautions, many of which largely take care of themselves. Thus the insane and the more serious cases of feeble-mindedness are segregated or, in some instances, rendered incapable of reproduction. But defectives are constantly being recruited as the result of the exaggeration in the offspring of characteristics in parents which have never led to social difficulty. This is especially true among psychasthenics and it is also true that among the offspring of such parentage may be found genius and unusual ability.

The best advice that can be given at the present time, therefore, seems to be against the mating of individuals in whom, or in whose families, psychasthenic traits or definite insanity are present. Unfortunately, this is often rendered difficult of achievement for the reason that such persons not infrequently are especially attractive to one another.

The education and protection of children presenting psychasthenic possibilities offers a much more hopeful field for prophylaxis. The traits indicative of this danger have already been outlined and a real psychiatric study (by which is not meant merely a psychometric examination) of school children, when generally carried out, offers a possibility of their detection. Once recognized, the next problem concerns the measures to be adopted. Here will have to be decided the advisability of removing the child from the mal-education in the home which will come from the examples of indecision and rumination in the affected parent or parents. Such measures may be very difficult to enforce and yet may be really vital. Segregation of the child among others similarly predisposed is for the same reasons unwise. The training that is needed is especially that of social association and practical activity. Group games and the development of such natural interests and aptitudes as the child possesses along lines of practical application are especially important. Especially careful instruction, given at the proper time and in the proper way, with regard to sex hygiene, a rational view of the true meaning and entire propriety of sex instincts, desires and happenings, the avoidance of suggestions which would encourage false modesty or prudery and the means of adequately meeting the demands of sex by directing the energies belonging to these functions into productive activity, artistic or practical or active games, must be paramount considerations.

It is not possible to say how much can be accomplished by these educational measures, undertaken at an early age, but it may be expected that they will fail to do more than ameliorate conditions in those individuals with severe degrees of constitutional deficiency, and it may be necessary in addition to prepare the child and the environment for a mode of life in which responsibility and demands for active participation are more or less restricted. Many of these people are highly intelligent and, since their interests are especially philosophic, it may be difficult to induce them to prepare themselves for a life work which is practical and not too responsible. It is therefore important to begin this attempt early and to train systematically. It is always possible to

permit the adoption of higher aims as these seem possible, but not so easy to relinquish them and consent to life on a lower plane.

TREATMENT OF THE DEVELOPED DISORDER.—In the treatment of the developed disorder it must be borne in mind that there is more or less constitutional deficiency. This it may be impossible to remedy even though particular symptoms may be successfully relieved. It is necessary therefore to consider the treatment of symptoms as well as the measures to be adopted in enabling the individual to live socially in spite of his handicap.

As a rule the patient first comes to the physician only when the disorder has become so obvious and so severe that he is more or less completely incapacitated for active participation in life. At this time it is not infrequent to find the suffering and distress so acute and disturbing that it is impossible to undertake any detailed investigation of the meaning of the various obsessions which can alone offer any real hope for their relief. At such time efforts must be directed towards tiding over the acute upset and awaiting a calmer period. For this purpose **removal from the home and isolation** from friends and relatives may be absolutely necessary. In addition **sedative measures**, such as **neutral packs** or prolonged **baths** and gentle **massage**, may be required. At times even drugs may be used and Janet recommends especially the use of **bromids**. As a rule a few days will suffice to bring about a subsidence of the acute agitation and it will be possible to enter upon a more definite plan of treatment. Where the acute manifestations of agitation do not exist this can be started at once.

The first step concerns the **establishment of confidential relations between patient and physician** and it is especially important, as Janet emphasizes, for the latter to show no evidence of doubt as to diagnosis. These patients often express the belief that no one understands the trouble from which they are suffering, that they are very unusual cases, no one ever suffered in this way before. It is usually easy to arrive at a diagnosis in a very few minutes, though a detailed examination for possible organic disease should never be omitted, and the physician can readily convince the patient that his case is not unusual by a few questions concerning symptoms as yet not described which experience tells him are likely to be present. In all subsequent dealings with the patient there must be no hesitation; everything must be met with calm assurance and the expression of perfect competence to meet any symptoms which may be declared.

The patient must be permitted and encouraged to give a detailed account of his obsessions and other symptoms with the object of securing a complete account as soon as possible and thus avoiding the necessity for constantly reverting to them. When this has been accomplished there comes the very important question of deciding whether an effort should be made to **analyze the symptoms** with the object of discovering their exact origin or whether it is better to **avoid stirring up old and painful memories** and merely endeavor to teach the patient to live in the present. The answer to this question is not always easy to give and

it must be realized that an unsuccessful attempt at analysis may leave the patient worse than he was before. The factors to be considered are the degree of constitutional defect, the age of the patient and the extent and duration of the symptoms. The more simple cases, that is to say those with but few obsessions, the younger individuals and those in whom the duration is not too great, offer the best prospects for successful treatment by analysis.

Analytical studies should not be lightly undertaken but, once started, it is especially important, yet often difficult, to know when to stop. The author prefers to do as little as possible and, once a definite conflict between instinctive and social self has been discovered which adequately explains the obsession, to avoid searching further for details and specific causes.

The method pursued in endeavoring to bring to light the "forgotten" episodes relating to the origin of obsessions varies with different physicians. Some use frank hypnosis which is, however, especially difficult to induce as the result of the indecision and doubt with failure to fully coöperate which are characteristic of the psychasthenic. Others use hypnoidal measures in which the situation is planned to impress the patient by semi-darkness, closed eyes, reclining posture, distraction of attention, etc. These will often serve materially to shorten the procedure and may be used. Personally the writer prefers to employ a purely conversational method, merely encouraging the patient by occasional questions and remarks to confide his thoughts freely. He is asked to speak out any thoughts which may arise in consciousness in association with the obsessive thought, feeling or act which is being studied. These thoughts must be pieced together and search made into any incidents thus brought up which may seem to have a bearing upon the actual evolution. The more closely the real conflicts are touched upon, the more evidence of feeling the patient will show and the more resistance will be encountered in obtaining a faithful relation of the incidents.

The bringing into consciousness of the forgotten incidents, or the realization of the conflict for which the symptom stands as a symbol,* is usually accompanied by a very decided feeling of relief and may, in the most simple cases, result in complete recovery. But far more often it serves only as a means of establishing for the patient a realization of the symbolic nature of the trouble, upon which may be built a constructive plan for meeting difficulties in the future. This means very often education in the physiologic nature of instinctive desires and feelings, similar to that advocated as a prophylactic measure. Upon this basis also it becomes possible to plan the future mode of life of the patient in such a way as to secure his coöperation. If he recognizes and understands his deficiencies or handicaps he is more liable to be willing to modify his life so as to avoid responsibilities and situations which may prove too great, and he has also a foundation for the establishment of modes of more directly facing situations in the future.

The selection of a suitable mode of life, with the educational meas-

* See under Pathology.

ures advocated, constitutes one of the most important points in the treatment directed towards meeting the constitutional deficiency which is present. It is not a matter which can be settled in a short time and may require prolonged effort and repeated trial and encouragement before a reasonable degree of comfort and success can be secured. This may extend over several years. At times it may even be necessary to retain the patient permanently under a parental type of control in which practically no responsibility is allowed.

As an extremely valuable adjunct to the educational treatment, reference should be made to the possibilities afforded by **occupational therapy**. This means training in steady application to some practical task which can be selected with due consideration for the natural interests and aptitudes of the patient and can be varied by alternations with amusements, games or different kinds of occupation. Through this means it may also be possible to determine more accurately the particular kinds of employment most suitable, the degree of attention of which the patient is capable and to establish hobbies which will afford an outlet for the instinctive needs of the patient in the midst of a more routine and monotonous employment.

But little has been said so far with regard to **general hygiene** and the **care of the many disturbances in body function** which may be so distressing. If it is deemed wise to undertake the analysis of these symptoms, which are symbols just like the other forms of obsession, very considerable relief may be secured. But if not, one must deal with them by assurance and quiet insistence upon the observation of general hygiene rules. Not infrequently these patients are much emaciated and they may be semi-starved. A prolonged period of rest and over-feeding similar to that recommended for neurasthenics is then indicated. Insomnia and general restlessness may occasionally demand attention and in severe cases may necessitate the use of bromid, but this should be avoided if possible and every effort made to correct the mental attitude which is responsible for the lack of sleep. Habit-forming drugs should be absolutely excluded and alcohol should never be used, although it often affords a very striking relief which is, however, quite temporary.

Prognosis.—The outlook for complete restoration to unrestricted social relationship is far from good. Improvements or, as they may be called, remissions are relatively frequent, but relapses, often with more serious degree of incapacity, are quite frequent. But while this general statement of the situation has some value, it cannot be applied quite indiscriminately to any individual case. There are many examples of disorder of this type which are quite favorable as regards recovery and it is therefore important to consider those features which seem to be of importance in determining the outcome.

First among these should be placed the influence of heredity or, in other words, the degree of constitutional deficiency present. The most unfavorable type of inheritance is certainly that of like kind. In estimating the presence of like inheritance, attention must be paid to the existence of psychasthenoid traits which may not have been sufficiently

severe to lead, or which because of the absence of serious difficulties to be faced have not led, to an actual breakdown. Many such people are described as merely "nervous." Often they have been simply negative characters, somewhat over-scrupulous, inefficient, timid, retiring and interested largely in the abstract. Pronounced insanities, especially the dementia præcox and paranoic types, are also of serious import. In close relation to this same question stands that of the age of onset. For, obviously, the greater the deficiency of endowment with energy, the greater the liability to break down under the lesser strains of the earlier and more sheltered years of life.

Another factor which has a similar bearing, but which is not so easy to determine, is the severity of the conditions of difficulty which brought about the breakdown. The outlook for recovery in persons who succumbed only under severe difficulty is far better than in those who have been unable to face the conflict between ordinary biologic needs and desires and social restriction which we must all inevitably face. It should also be obvious that the more impossible it is for the individual to adjust to the simpler difficulties of life, the wider will be the evidences of such deficiency. Hence the more widespread and varied the obsessions and their associations, the worse the prognosis.

Psychasthenic reactions which have been practiced frequently and over long periods also tend to become more and more settled habits with the consequence that the chances of recovery become reduced with the prolongation of the disorder.

The prognosis as regards life is essentially good and sufficient consideration has already been given to the dangers from impulsive acts and complications. The possibility of the development of the disorder into a true insanity, in the sense that the patient loses his grasp of the real facts and confuses his obsessions with realities, is practically negligible although there have been dissenting opinions. Nevertheless, a very small minority of the severe types of this disorder do require permanent isolation from society, because they are utterly unable to provide, or even care, for themselves and by their crises of agitation and ruminative acts may seriously disturb and interfere with others. At times the only available place for such isolation is the hospital for the insane. Here they also are often better understood and more tactfully met and considered so that, miserable as the condition of these unfortunates is, they may be less unhappy in such an environment with its relative freedom from responsibility and social demand than in their homes. Such a termination is rare and the adoption of this plan of treatment must be considered absolutely as a last resort.

Pathology.—Psychasthenia presents no known morbid anatomy. The pathogenesis is extremely vague and indefinite. The facts of heredity and age of onset do, however, tend to suggest the existence of some deficiencies in structure, the nature of which is quite unknown. Janet in analyzing his most admirable exposition of the facts of the disorder comes to the conclusion that the essence of it consists in a prolonged "lowering of psychic tension" with defective grasp of reality. He also

points out that various means which stimulate cerebral activity will result in considerable amelioration of symptoms, unfortunately only temporary. Thus alcohol, febrile intoxications, absorption of attention and sthenic emotion will all produce this effect.

These considerations, accurate as they are, offer, however, no real explanation for the lowering of tension, although he suggests that any condition which results in enfeeblement of the organism, such as disease, fatigue and depressive emotion, may precipitate the disorder. This simply leads us back to the conception of some deficiency in structure, often inherited, but possibly emphasized by damage suffered during life at any time after fertilization of the ovum. Unsatisfactory as this conclusion is, it is impossible at present to be more precise.

But the situation is different with regard to the symptoms of the disorder. The mechanism of their development, granted the initial deficiency in vigor of action, is far more readily intelligible. The various symptoms are nothing but symbols substituted for the real facts of situations in which the individual has been unable to reach a satisfactory conclusion. To avoid the difficulties, the real facts are "forgotten" and some more or less accidental association is adopted in consciousness in their place and becomes endowed with the attention qualities which belonged to the original situation.

Donley has given some excellent illustrations of obsessions of quite simple type and of restricted extent, in which this mechanism of substitution is well shown. Thus he quotes the case of a man who, without other manifestations, had been for fourteen years absolutely unable to get upon a street car bearing an odd number. Under hypnosis it was learned that the difficulty dated from an occasion upon which he saw a child struck and injured by a street car. He happened to notice that the car bore the number 213. In the future a car bearing an odd number re-arouses the affect of fear and horror but the actual incident has been forgotten or dissociated. The generic "odd number" has even been substituted for the more specific 13 which is too closely associated with the actual incidents of the situation.

A much more complex illustration is afforded by a case of the author's in which, among numerous other obsessions and fears, there was a constantly recurring mental rumination evidenced by innumerable repetitions of the acts of dressing and undressing, often kept up incessantly for hours at a time. From the patient it was learned, after prolonged study without hypnosis, that the beginning of this symptom dated from an experience of girlhood. At a vaudeville show she witnessed the performance of a professional hypnotist whom she described as a very large man, in white tights. She was deeply shocked by the dominance of the genitalia, produced by the nature of his apparel. In speaking of the performer before the show, a girl friend had enlarged upon the will power possessed by this man, and had said that if he wished he could even make you undress and go out naked. Afterwards the patient began, symbolically, to submit herself to male domination by carrying out a part of the act suggested by the girl friend. The true

meaning, however, was probably never consciously grasped and the incident itself had been "forgotten," only the chance association being used in the future as a means of expressing desires which the patient was unable consciously to face. In this case it is easy to see that the rumination is a source of gratification and to understand why the patient persists with it in spite of the discomfort and disability which result.

This type of mechanism is present in all the different manifestations whether fears, obsessive ideas, acts or tics. All of them express an effort to escape from reaching a full grasp of, and definitely deciding, some situation of difficulty. These are most often situations demanding that the patient cease to be a child dependent upon the authoritative direction of others and assume individual responsibility. Difficult situations are those in which there is a conflict between individual desires and social restriction and to which belong, therefore, strong feelings. The psychasthenic, as we have assumed, is, for some reason, lacking in the ability for the decisive reaction which strong feeling implies. When such reaction is called for he sidesteps by forgetting the real facts and adopts a symbol instead. It is therefore not surprising that many of the manifestations must, in ultimate analysis, be referred back to sex feelings, for these are not only of utmost importance in the maintenance of life, but are also the activities which are most severely repressed by social requirement. That they are not necessarily sexual in origin, however, is amply illustrated by the analysis of Donley quoted above and of many other writers.

The particular form, or content, of the obsession is largely the result of accidental association. But it is probable that the principle of overdetermination suggested by Freud plays a considerable part. According to this conception, associations which have more than one point of contact with the dissociated experience are more likely to be adopted than others. Thus, in the illustrations given, there is a very frequent, popular belief in the unluckiness of the number 13 which would be an additional bond between misfortune and the car number besides the purely accidental one of this particular car having caused the accident. In the case of the girl patient, besides the chance remark of the friend, there is an obvious association between the ruminative act and the sex experience which had been dissociated. This mechanism explains why it is that identical symbols are often selected, under quite different conditions perhaps, by many different persons.

Historical Summary and Distribution.—Although the titles "neurasthenia" and "psychasthenia" are of quite recent origin, the former having been devised and first used by George M. Beard, of New York, in 1868, and the latter by Pierre Janet, of Paris, in 1898, it must not be supposed that the conditions included under these names had not been clearly recognized and described long before. Excellent accounts of the history of functional disorders are given by several authors, notably by Arndt and Dornblüth. According to the former the earliest effort at a real explanation is to be found in a work by Fernel, entitled "De

Abditis Rerum Causis," published in 1540, wherein is described "nervousness" arising from "vapors" resulting from retained semen or menstrual blood.

In 1765 the Scotch physician Robert Whytt published a book on "Disorders Commonly Called Nervous, Hypochondriac and Hysterie," in which he quite clearly outlines neurasthenic and psychasthenic cases and even warns against the indiscriminate use of such terms when he says: "Physicians have bestowed the character of nervous on all those disorders whose nature and causes they were ignorant of." He gives as predisposing causes for these disorders: "(1) a too great delicacy and sensibility of the whole nervous system; (2) an uncommon weakness or a depraved or unnatural feeling in some of the organs of the body." He further points out that the delicacy and sensibility of the nervous system "may be either natural . . . or produced by such diseases, or irregularities in living, as weaken the whole body especially the nerves. Long or continued fevers, profuse hemorrhages, great fatigue, excessive or long continued grief, luxurious living and want of exercise." These opinions are worth quoting for the reason that they cover very exactly the views which have since obtained.

In 1838, Esquirol described many of the disorders grouped here under psychasthenia with the title of "monomanie raisonnée." During the early part of the nineteenth century also a number of works appeared upon "spinal irritation" following traumatism which closely corresponds with what has since been described as spinal neurasthenia. In 1865, Austin Flint, in "Principles and Practice of Medicine," devoted part of a chapter to "nervous asthenia" as denoting "a morbid condition sufficiently common in this country, especially in the larger cities. The propriety of considering this a functional affection, and the name have been suggested by my colleague, Prof. Fordyce Barker. As the name signifies, debility or prostration, affecting especially the nervous system, constitutes the affection."

The first real generalization, however, came with the work of Beard: "A Practical Treatise on Nervous Exhaustion (Neurasthenia)," published in 1880. In this book, as in many later treatises by other authors, no distinction is made between neurasthenia and psychasthenia. Beard, who gives full credit to Flint, believed the disorder to be especially an American disease but in the later editions acknowledges that it is widespread over the civilized world. He was especially grateful to Erb, who adopted the name "neurasthenia spinalis," as a substitute for spinal irritation. Gradually Beard's concept has been accepted and adopted throughout the world, though many, especially of the older neurologists, protested against its use as a cloak for ignorance, much as Whytt had done in 1765.

Janet used the term "psychasthenia" in "*Névroses et idées fixes*," published in 1898, and followed this by his epoch-making work "*Les obsessions et la psychasthénie*" in 1903. He includes neurasthenia as a special subdivision.

The widespread extent of these disorders is testified to by the many

monographs in various languages which have appeared since the middle of the nineteenth century. Unfortunately there has been no general agreement upon definitions and these disorders are often included with hysteria under the general heading of Psychoneuroses. In general it may be said that neurasthenia and psychasthenia are common in all civilized communities and that while more frequent among brain workers and in large cities, they are not by any means limited to them. Thus Belbèze has found them widely prevalent in a large and strictly rural and rather primitive community.

We have at present no means of estimating their frequency, but it is certainly very great. Many neurologists have stated that such cases make up a large proportion of their practice and there are unquestionably many more in the clientèle of the various specialists.

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CHAPTER XVII

THE HYSTERIA GROUP

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Definition.—There was never a time in the history of medicine when there existed so great a tendency to discard static and limiting definitions for a closer actual survey and interpretation of clinical data. This is due chiefly to the fact that the genius for nomenclature has overreached itself. The human intellect demands safe and sure ground on which to stand and from which to work, and for this reason it tends to fix its concepts. This is practical and useful so long as intellect is not self-deceived and does not utilize such terms to stabilize concepts which ought to continue to function as alterable hypotheses for continued investigation. At the present time modern thought based upon experience and intensive research tends to outrun all older concepts of hysteria which have become thus fixed.

The term *hysteria* has stood for useful concepts varying through many ages and many forms. This has meant that the formulation of the precise significance of hysteria to any age of medicine served a purpose at that time which later gave way to some other. This indeed has been the history of the term. And yet there has always been present the danger of limiting thought and therapeutic practice by definition and by an idolatrous adherence to it. For from this characteristic of the human mind the scientist is by no means exempt. The gradual turning over of the concept and definition of hysteria has, however, gone on steadily, responding to the equally inherent tendency in the human mind to progress and expand its former limitations. Fortunately this also is not foreign to the practical scientist.

To-day the progress of medical thought and effort is imbued with the realization of the continuous striving of energy to find expression and to unfold itself in devious channels, and it conceives of the blocking of energy as the cause of suffering and disease. For this reason all groups of manifestations of such disturbance of function have to be considered far more broadly and deeply than the confines of any terminology. This energy creates its own hindrances because it tends to make its way

expressed at once through such need and the set toward its gratification. When, therefore, the wish, or rather the expression of it, is blocked by external circumstances or by the inability to make such an adjustment that the energy expression shall be brought into healthy and free relation to the outside requirements, the result is twofold. In the first place there is psychic and automatic discomfort and secondly there is an attempt, even though an imperfect one, at a readjustment which, however, aims to lessen the original psychic and physical burden. This leads to the creation of the many symptoms which are the outcome of such disturbance and imperfect readjustment of function. This may occur, as Kempf points out, when the individual is "clearly-to-vaguely conscious of the nature and effect upon himself of his ungratifiable cravings," that is, when he exercises upon them merely a suppression from clear consciousness. The various forms of energy striving or wish which cause such experience he has named under the "shrinking from responsibility" or "the liability which the presence of strong affective-cravings would occasion were they exercised; a dread of anything which might reveal organic inferiority or lack of power to effect the ends of the real world; with these the definite fears of loss of external things as money, or of honor or freedom, also of receiving pain or injury. There is also a fear of not meeting the expected requirements which tradition and affection have laid down. Oftentimes, too, there is a positive desire toward some perverse object unpermitted by society or toward an unattainable or unresponsive object, or hate, shame, and disgust for certain objects. This mild suppression of these tendencies results in hypertensions or hypotensions of the various autonomic (visceral) portions or segments of the body and the long line of functional disorders so commonly seen comes into play. Some of these appear to the patient as mental phenomena, such as distressing dreams, reproach, persistent thoughts which will not be dismissed or neutralized. Others tend to register themselves at least so far in bodily sensation that they appear as a decrease of power to coördinate the thoughts, inability to learn or to concentrate, to use the "brain" according to common conscious complaint. Or there follows the long line of more distinctly physical disorders, often, of course, associated with the foregoing. These grade from what may still be interpreted as errors or accidents to headache, dizziness, stiffness or weakness of various muscles, interference with secretion of mouth and stomach glands, dyspnea, tachycardia, dysmenorrhea and amenorrhea, sexual impotence, hyperirritability of structures otherwise diseased.

In this "*suppression group*" belong those disorders which the war has brought so prominently to the fore and which have been very fully described by the French writers. They have manifested themselves in what have been called the reflex nervous disturbances or designated and described more in detail as various contractures, paralyses and paretic states affecting one or more members or parts of members, muscular atrophy, hypotonus and hyperexcitability of the muscles, changes in skin and tendon reflexes, change in excitability of the nerves, dis-

urbs of sensation, thermic disturbance and vasomotor disturbance, cerebry and trophic disorders of various sorts. Such physical findings are excluded from hysteria according, for example, to Babinski's nomenclature, who reserves the term hysteria for those manifestations which he somewhat obscurely states may be caused by suggestion and cured by suggestion. To these latter he gives the more distinctive name of *pithiatism*.

Etiology.—The more inclusive dynamic conception, however, which follows the development led on from Janet by Freud, for the sake of a workable etiological exploration as well as of a practical therapy, would bring these manifestations back into the broader and deeper concept of hysteria. Etiology, then, becomes a matter leading into a deeper terrain, out of which both the more strictly mental condition which Babinski defines under *pithiatism* and these apparently more purely physiological manifestations which he calls reflex disorders are, after all, animated alike by the unconscious reaction to war conditions, including the effect of slight trauma, which Kempf outlines in his description of the effects of *suppression*. The difficulties presented to these cases in warfare, increased and precipitated by special circumstance, a wound, a sudden overwhelming threat of injury, as in an exploding shell, with perhaps temporary physical "commotion," form the final cause why the fatigued personality no longer reacts vigorously toward the outside environment and retreats into pathways of less effectual activity. For these at the same time serve the unconscious or barely conscious ends, such as, for example, a desire for release from the extreme difficulties of warfare, consciously unknown and unacknowledged to the individual. This reaction may activate any of the pathways over which the wish may bring a direct or indirect fulfillment. The roads to fulfillment might be those ordinarily called voluntary ones which may interfere with certain ordinary voluntary functions, but they can just as well, by the same mechanism of mental stimulus of any physiological pathway, find their way into expression through the reflex arcs controlling tendons, skin, heart, vasomotor system, trophic conditions of skin, hair, nails, etc., or by those which adjust muscular states. In short the patients have consciously or unconsciously the whole range of bodily activity, sensorimotor or autonomic, at their disposal. The reflex vegetative arc has not been sufficiently taken into account, although this is possibly older in development than the more easily recognizable sensorimotor reflex arc and its response, and even the often observed reactions of the latter type really take place over such arcs. When research has brought to light more facts in regard to the establishment of such arcs and their continued function in autonomic activities, the concept of hysterical conversion will tread on surer ground and its mechanisms will be seen under greater illumination.

The foregoing conception of hysteria is made still more logical by Kempf's pursuance of the idea of *repression*. This works in the same fashion as the suppression but with a more profound effect upon the individual's personality. For the repression signifies that the individual

tries "to prevent the autonomic cravings from causing him to be conscious of their nature or needs and influence upon his personality. He succeeds in this by maintaining a vigorous, incessant, defensive coördination ('concentration of attention') of his *egoistic* wishes upon a course that compromises, as a resultant of converging forces, with the repressed cravings." Here the individual not only prevents the affect which represents his craving from passing over into unacceptable form of action, but he also does not permit himself to be conscious of its existence or the needs it expresses. As far as consciousness goes he forgets it. In the mere suppression, on the other hand, he *permits himself awareness* of the needs for which the affect stands but *prevents the expression* of those needs in overt acts.

The symptoms which such a state of affairs causes can be seen as far as hysteria is concerned in sensory disturbances, anesthetics, specific, localized and general, hyperesthesias and paresthesias and in cravings for certain forms of stimuli, perhaps sexual, perhaps esthetic, often for painful or semi-distressing ones—masochistic. There may be amnesias and misinterpretations according to the buried affect and misrepresentations. Convulsions may occur but without loss of consciousness. There will be interference with segmental functions, incoördinations, spastic and postural tensions with also simulations of postures. There will be increase of muscular tension, of blood-pressure, of exophthalmic tensions with hyperactive functioning of the glands of internal secretion, or there may be interference with these functional activities with lowering of their tone and tension, according to the form of unconscious attitude. For this may be that of a compensatory striving to discharge the concealed emotion or of a regressive tendency which tends to draw the personality further back into asocial aim and wish reaction, that is, further away from reality into the realm of phantasy production and activity.

These physiological reactions may be accompanied by or substituted for, in a large degree, by corresponding mental attitudes, overcompensatory in nature, such as excessive zeal in reform or in good works, sense of inspiration and exaltation of mission, exhibitionistic tendencies in dress, voice, manner of behavior, or they may represent the regressive attitude with evasion of responsibility, indifference, inefficiency, absorption in childish dreams. There may even be marked *dissociation* conditions manifest in definite and discrete sensory disturbances, in an un-governed exercise of excretory functions, more or less plainly erotic in character. There will be an uncontrolled or a more systematic striving to compensate, there will be apprehension, panic over loss of self-control, anger and rage as an unacknowledged defense against the affective cravings, impulsive action, postural effects and also phantasies as a substitute for denied reality. There may be also severe visceral distress and motor derangements. These patients show dreamy deliria.

HISTORICAL.—It is from the grouping of the earlier hypotheses of hysteria that this view of the etiology and this outlining of the hysterical manifestations has gradually been worked out. Hysteria has been dis-

extended from the earliest days of formal medicine. It even entered largely as it could be expected into prehistoric legends and folk lore and it can be traced in written literature of all times. It formed the subject matter for lyric and drama, and was represented in plastic and graphic art. It was recorded by historians and chroniclers of events and entered into the consideration of the law makers and, of course, played largely in the religious world. From ancient times there was the conception of unsatisfied longing, which was connected with the uterus. This, like so many once intuitively held, afterward intellectually discarded, ideas, returns again in the newer intellectual conception which ascribes to functional and organic disturbance an emotional psychogenic cause in unsatisfied emotional striving, particularly in its most intense and most repressed form, the reproductive instinct.

The uterine theory of hysteria has, of course, had its exponents up to the present day in the more anatomical form through a consideration of nervous connections. The extension of the idea of hysteria to the nerves as the source was a natural development and was closely associated with the humoral doctrine which also still has its remnants in medicine. Boerhave, for example, attributed hysteria to the effect of an intoxication of the abdominal nerves which resulted from the stagnating or obstructing action of the humors of the body. Such manifestations as cutaneous anesthesia, deafness, blindness, hysterical aphonia and tremors were described as early as the seventeenth century, and these were attributed to the head instead of to the uterus or the intestines.

Present-day conceptions of hysteria were, however, put into form by the Charcot school, when the various hypotheses, the various ways of approach and the various aspects in which hysteria was conceived and defined began to be brought more consistently together. The underlying spirit which governed this development prepared the way for the recognition of the dynamic element which belongs to the mental life and which therefore permits of the recognition of the psychogenic meaning of hysteria and a pursuit of its mysteries along this line. Since Charcot's day the approach to hysteria whether from the more purely psychological standpoint, the physiological or the biological has made more and more use of the psychogenic idea and come to a greater comprehension of its extent and significance. At the same time that there is a more unified and comprehensive conception there is also a tendency to dismember the large conglomerate of phenomena in order to give them more intelligent and effective therapeutic consideration.

Charcot laid emphasis upon the idea of *dissociated personality*, the psychological disturbance and upheaval which marked hysteria. This was elaborated further by Janet, who also tended to dismember the larger group still further under this aspect and approach it in a more analytical form. The factor of suggestion as a causative element and as a means for cure entered largely and was developed from this point of view. Other followers of Charcot pursued certain other definitions. Möbius included the ideational source of morbid phenomena to cover physical as well as psychical manifestations, all having a common

psychogenic origin. Briquet and Gilles de la Tourette have presented much in the way of description and discussion of the manifold symptomatology.

Babinski has rendered special service in the dismemberment which naturally and necessarily followed upon the comprehensiveness of Charcot's studies. But these tended again to make a fixed concept as did the older more partial concepts, which had to be broken into for the flexible purposes of practical investigation and therapy.

He has done much in developing the idea of suggestion or persuasion, to which he applies the term pithiatism. He has also laid stress, as has been mentioned, upon the "physiopathic" symptoms which he prefers to split off from hysteria into the disorders of reflex origin, excluding these from the psychogenic forms.

He has thus given the necessary emphasis to the actual physical phenomena which appear to be associated with reflex disorders; but in separating them off from the hysterical group has not only left them unsatisfactorily explained as he himself acknowledges, but he would thus prevent their explanation through a possible "conversion" mechanism which operates on a lower, more basic nervous level than that of the more obvious sensorimotor reflexes. This is that of the vegetative or visceral reflexes already mentioned. Nevertheless he has done much toward giving more definiteness to the hysterical grouping by removing much diagnostic error. Too many somatic disorders due to primary physical or chemical causes had heretofore been brought under this one vague explanation, and also certain trivially entertained ideas needed the more definite and precise form of definition which he gave.

The question of simulation and deceit has been looked into by Babinski but without promising results. This much discussed question receives its best illumination and a more practical possibility of approach from the fuller conceptions of hysteria based upon the more analytic consideration of unconscious psychogenic factors. It will be further taken up in connection with these.

Other writers have dwelt upon the physiological side of hysteria. The earlier English school tended toward this, and Sollier and Binswanger on the Continent have given such definition in physiological terms to the psychological processes. This is spoken of in terms of functional disturbance of the cerebral centers, which then manifests itself in the varieties of disturbance according to the centers disturbed, vasomotor, trophic, visceral, sensory, motor and psychic also. The correlation between the psychic and physical processes is therefore interfered with so that the disturbance manifests itself in both directions. The careful experimental and clinical work of later investigators tends to give such a correlation of mental and physical factors much greater precision and clearness in definite physiological and psychological terms. Both are better explained in terms of the personal craving, wish dynamism, translated into terms of the visceral cravings and in turn intensified or relieved as the physiological activity satisfies, restrains or aggravates these cravings or wishes. The physiological views which

tend to explain hysteria upon the basis of the reflex action of localized centers lie also in the field of this close relation of mind and body through the vegetative innervation, though they have tended to separate off this form of explanation into two separate compartments. This, for example, is the attempt to explain hysteria as due to localized or general vascular changes in the brain or to some other effect chiefly of a vasomotor instability throughout the body.

MODERN PSYCHOLOGICAL CONCEPTIONS.—Such are the hypotheses which represent the growing attention given to the conception of hysteria. All this has been widely and at the same time intensively studied by Pierre Janet and, after him, by Freud. Janet pressed on into the region of the subconscious where he believed groups of ideas to exist which are barely known or quite unknown to the ordinary awareness of the patient. And yet, according to him, such ideas are active there, having the ability to invade the personality largely in a negative way in that they divert what would otherwise be the conscious attention from its external tasks or interests and so seem to cut these out of the field of consciousness. This peculiarity Janet saw as disorder of attention or a narrowing of the conscious field. The result of this is a splitting or disintegration of the personality, the elements which ordinarily form by their association the whole personality being retracted from this field and the various elements or systems of elements being able now to function independently in the individual. Janet conceived of the hysterical state as being synonymous with the hypnotic state and with suggestibility as the prominent characteristic of either state.

The followers of Janet have continued their pursuit of the dissociation theory with greater emphasis laid upon the etiological features which caused such dissociation and with the effort to bring the personality back to its healthy synthesis based upon such causal factors. Freud particularly has pressed deeply and determinedly into the separate etiological factors and given very careful investigation to the mental processes and mechanisms through which the dissociation has been brought about and through which it maintains itself. His investigations, stimulated first by his work with Charcot and Janet, were carried on in association with Breuer, who also pursued his investigations in this direction. But Freud as the younger man struck out more definitely into the newer pathway and all that it involved. For his early experience led him deeply into the most intimate and intricate mechanisms of the mental life. This had been suggested also by the testimony of older medical men, who, however, did not practically press their theoretical convictions, as he felt bound to do. Breuer had already stated the belief that psychical or physical trauma lay at the basis of the hysterical symptoms and as Freud developed this idea he was led at first to give somewhat exclusive place to genital trauma or sexual trauma in the narrower sense. Later, however, he extended this conception to exclude the idea of a necessary actual genital trauma and also widened the concept sexual to include all manifestation of the instinct of race propagation or reproduction, with all forms of production or reproduction that grow out

from this and all forms of personal reaction which are associated with the love life and its wider extension into society.

Just here the interpretation entered in, thus helping to explain why the disassociation discussed by Janet had taken place. By thus finding the reason for such a result in the personality and the mechanisms by which it had come to pass, Freud believed himself enabled to reëducate the patient back to health, in a sense by a reversal of the process or a following of these mechanisms through analysis into the structure which they had built up and into the material out of which and by content of which they had been able to construct the morbid reaction. This he attempted first by the method of hypnosis, classical at the time. He replaced this technic later by the conscious rational coöperation of his patients, which he called psychoanalysis, believing that in this way the morbid process and the process of reëducation could be more effectually and lastingly pursued with more rationally established and lasting results.

For Freud held that the most important mechanism upon which all the others depends was that of repression. This is the passively directed mental factor which tends to put out of mind all that is unbearable, all that would interfere with the individual's social position, his own moral or higher personal estimate of himself. All this repressed material, in its direct form, is of an unacceptable nature because it concerns chiefly the individual affective needs which naturally and instinctively tend to gratification in individualistic ways and often in conflict with social demands. Gradually Freud came to lay more and more emphasis upon the nature of the whole personality which was unable to choose and form those reactions which would make a workable compromise between the individual cravings and the social position. This led him into a fuller study of the mechanisms at work and the entire mental groundwork in which they operated. The content then of the repressed material, or the split off complexes, was found to be that of this instinctive material of personal craving, for which Kempf has utilized the term *autonomic* cravings. The various forms which this material assumes are described in the forms of individual wish and the secondary fear, hate, disgust and the like which have been built up against these. They may still be subsumed under Freud's conception of sexual desire if there is kept in mind the broader conception with which Freud uses this term. In fact, it seems necessary to do this, although perhaps the term sexual is unfortunately misleading, as it tends to be too narrowly interpreted.

It was Freud's desire, however, based upon his earnest conviction to which his work had brought him, to keep always in mind the reasons for the repression and for the difficulties of keeping the repressed elements in the synthesis of the personality. Therefore, he was led through his continued investigations to the very source of the personality, the unfolding of the libido or creative energy. He saw the need for libido expression through the various channels of the personality and recognized that the fundamental form of such libido expression lay, strictly speaking, in the sex life. Here it has been longest in activity,

biologically considered, and so it has all the force and insistence of a primeval function ineradicably imbedded in the very human constitution. Life is passed on by this function and there would have been no evolution, in living matter, without it. Nevertheless, because of its force and its persistence it has necessitated a greater control than any other form of impulse, has in fact made necessary a strong repression. This has been in the service of a diversion of the energy and desire represented, the libido, into other channels, which has had for result the widening of the personality as regards the individual and the building up of the complexity and extent of civilization as regards the entire race. Yet as no human process is carried out perfectly, this process in the interests of development, individual and racial, has manifested all degrees of incompleteness and carried with it grave dangers.

No individual has yet acquired a perfect capacity for such repression and for the transformation of impulse and the adjustment of restrained desire which this implies. Some individuals develop less of this capacity than others. This it may be conceded may be due to greater constitutional differences. It rests also largely, often in conjunction with the former, upon imperfect education in childhood. Education here is used in its widest sense to mean the bringing together into one unified and progressive purpose the trends, tendencies, needs, wishes of the individual, so that the personal wishes, conscious and unconscious, are sufficiently gratified while the social relationships are also made satisfactory. But here may result the splitting off of individual factors, which because they are difficult to manage and bring into line, assume the aspect of inacceptability, frightfulness, or what not.

All this Freud studied largely in the field of child psychical development with its many interests serving the as yet obscure but already present reproductive instinct. The self-preservative instinct is involved as well, but manifests itself chiefly in the realm of the greater instinct, that of reproduction. This because of its necessarily greater capacity for biological and social service has the more possibility for diversion into many forms and channels and at the same time the greater possibility of dangerous deviation upon unproductive channels and the greater danger of repression in consequence.

SEX THEORIES.—In this way Freud found that the hysterical disturbance was the result of disturbance of the reproductive instinct. It is necessary to consider this in its narrower sense as well as the wider because of the difficulty, from the view of individual and racial need, of training the definite individual sex need to its healthy satisfactory function. This lies in a gradually developing pursuit of objects and interests in the external world until the adult love object is found and an adjustment of mutual progress and workableness is established. Along with this or preceding it the gradual development of functional interests pertaining to the body should go into a canalization of supreme adult interest in the genital maturity and function. At the same time with this specialized line of development the individual attains an accompanying maturity of development of interest and activity in broader social chan-

nels which make up usefulness and satisfaction and also make for healthy coöperative physiological functioning through all the channels of the body.

It is just this broad psychosexual development which Freud finds interfered with and this interference which he makes the basis of those special manifestations which are termed hysteria. Already in infancy the proportional relation of these various forms of manifestation and interest, individually and socially, physiologically and psychically, is disturbed and sexual activities and psychic attitudes develop, which, if they continued to be manifested in the period of adult life, would be called perversities. They are, however, for biological reasons, reproductive and self-preservative, driven out of the conscious life of the individual and refused this manifestation so far as the physiological personality is concerned. Psychically also, that is in their mental interpretation on the part of the patient, they must not be recognized nor expressed in their direct form. Therefore, in the first place, these autonomic or physiological needs and their purely psychic interpretation are kept forcibly out of sight. Here, in the second place, they are the prey of the unconscious phantasy which dwells upon them seeking in dreams to obtain the gratification which reality denies. These are unconscious dreams, but often in hysteria pass over into conscious ones. They so transform and distort as well as magnify the original needs or cravings, as well as the form in which they may escape the repression and get out into some form of expression, that there arises all the protean symptomatology of hysteria. These may then exist in the phantastic dreams, erotic longings, delirium, etc., beside all the train of anesthetics, paresthesias, trophic disturbances, in fact all the extensive variety of physiological manifestation to which attention has already been called.

This view of hysteria may be summed up as that of initial repression, followed by dissociation and then the conversion of this material into symptoms. There should be noted in regard to this mechanism for the formation of hysterical symptoms that which Freud has insisted upon in regard to the mechanism underlying any form of psychoneurosis. It is that these mechanisms belong to the equipment of the mental and physiological life of every individual. There must always be conflict between the elemental individual tendencies and the needs of that individual dependent upon social adjustment. It follows then that repression is always useful and to some extent dissociation and conversion, but it is the degree of these and the use to which the personality puts the repressed material, the form and content of the complexes constructed from it and the way these are handled which constitute reactions that are hysterical or otherwise psychoneurotic or that serve the individual well and healthfully.

The hysterical reaction is to split the material and the complexes off and permit them to function apart, while a healthy use of repression profits by the amount of pressure brought to bear in the personality by the force of the repressed material to make a new and more effective adjustment of the energy contained in the unconscious or semiconscious

depress. In the pathological situation there is inability to readjust effectively by bringing about always a new state of affairs in accord with a continuously changing reality instead of the unacceptable form of action or the one which would cause the individual too severe a deprivation. In other words in the pathological state there is inability for making the healthy compromise by which the individual must continually establish and reestablish his relation to society.

This healthy compromise necessitates the attainment of a level of conduct where the conflicting tendencies and needs can be brought into a workable synthesis, but the hysterical character has not learned to reach such a higher level with the *whole nature*. Certain tendencies remain in the infantile form while there is a compensatory striving often on the part of the social side of the nature to maintain a very high level. Thus the breach in the personality tends to widen rather than otherwise and the possibility of synthesis is more and more displaced by the dissociated form of activity. In order then to neutralize the psychic distress occasioned by such a split in the personality an artificial synthesis has to be made which is not too strictly in accord with the laws of external reality. Other forms of neurosis attain this end in various ways which still disguise the actual content of the complexes from the individual. The peculiarity of the hysterical mechanism which effects this is that it utilizes the pathways of bodily innervation for physically unsocial ends or those which pathologically compromise with usefulness and the force of the affect is diverted into the various bodily channels indicated. This force manifests itself in the variety of somatic disturbances, "converted" into these phenomena, and the psychic weight, pain, of the conflict is lessened.

Symptomatology.—The history of the concept of hysteria as it has made its way through the history of medicine up to the present time necessarily loses itself in the consideration of its etiology. The latter, however, may be considered from other angles, which serve to bring out more particularly the relation of hysteria to other disease manifestations, the particular forms in which it can present itself, its relations to the various levels of nervous activity, and therefore its varieties of somatic manifestations. All this points the way toward its treatment and prophylaxis.

Enough has perhaps been said to mark the two distinguishing features of the modern approach to this problem—that of the dynamic or energy point of view which gathers the whole topic for any individual patient into a comprehensive search for the character and general aim of the personality, and the analytic point of view which seeks to determine very definitely the fate of this general aim and direction of energy striving as it has become split and broken into ineffectual fragments. The latter view seeks to eliminate a faulty diagnosis which would lazily overlook certain symptoms or confuse under the term hysteria certain definite symptoms which really mark organic lesion of various sorts. The comprehensive dynamic point of view equally forbids a careless indefinite diagnosis, for each symptom, as an expression of en-

ergy striving, takes on a new significance and a new importance in the expression of the personality. No symptom grouping, no matter what it may be, can be effectually understood or conscientiously treated without a careful pursuit of each one of the definite features in its origin and meaning.

Careful sifting has resulted from this precise attitude and also from the more exact and intensive methods of diagnosis which physical science makes ever more possible. The more exact methods, for example, of blood examination, have enabled the physician to lay his finger definitely upon the dividing line between actual toxic disturbance which produces thermal disorder, once attributed to a vaguely conceived hysterical reaction, and the strictly psychic cause of the disturbance. A great deal of such vague falsity has been eliminated from diagnosis of disturbances of the nervous system, which now can be clearly detected to be of physical nature. And, on the other hand, this more careful distinction between organic lesion and purely psychogenic disorder does not interfere with the recognition of the hysterical conversion but gives to it its proper relation to any organic lesion that may exist, and recognizes the hysterical tendency to exaggerate any such lesion or utilize such a pathway of least resistance for its activity. In this way the hysterical character may manifest itself and play a no small part in weakening resistance to an invading infection, or in aggravating an already started disease process, lessening the chances of overcoming the disability and even precipitating a serious result, fatal or otherwise.

VEGETATIVE SYMPTOMS.—This close connection between the psychic impulse and its reaction, that is, between the psychogenic, sensorimotor and the vegetative is coming more under the observation of the neurologist, through the knowledge which is slowly being gained of the action of the autonomic or vegetative nervous system, particularly as it is operative through the glands of internal secretion. Here through the lowest reflex arcs and through the influence of the internal secretions, a wide range of somatic activity is put at the disposal of the psychic wish and as this portion of nervous activity is unconsciously activated, it forms a vast mechanism particularly at the disposal of the unconscious phantasies in their effort to obtain the satisfaction which the individual is incapable of attaining upon the higher conscious levels.

In this way it can be seen how the field is sufficiently extended to admit those reactions which have been named the disorders of reflex origin and the wide range of vasomotor, trophic disturbances, disturbances of tonus, of sensation and the like. It can easily be seen also how these fall beyond the range of ordinary conscious "suggestion" or "persuasion" for they are activated so far beneath the conscious surface. This brings them, however, no less within the range of mental therapy and emphasizes the need of a deeper analytical penetration, until deeply unconscious motives shall be brought to light which have the power to activate these deeply unconscious pathways. Not that the simple suppression of affect as in some of the war neuroses may not utilize these somatic pathways also which stand at the service of unconscious de-

sires. For experience with war neuroses has testified that these disorders of function were in some cases reached by a superficial analysis or perhaps through a diversion by more indirect mechanical means of the energy infusion of these pathways so that the normal useful pathways were once more chosen and the ordinary functional activity was no longer interfered with. Yet even in such a case one must be sure that the cure has not been a merely apparent and superficial one, allowing the energy transference to be made to some other, still more concealed pathway. For then the hysterical reaction may reappear at a later period, in a still more pernicious, somatic, or some more psychotic form of mental reaction. For this reason Freud has advocated the exhaustive analysis in order not only that individual symptoms may be followed up but that their cause be followed into the wish content. After such analysis the patient is enabled consciously and intelligently to recognize the real condition so that the choice of reaction may come within his conscious power. Clear intelligence governing at last emotional reactions he may then utilize those sensory and motor pathways over which he can fulfill the wish in a way compatible with his ethical and social desires and at the same time with his most complete organic functioning and health.

SIMULATION.—The question of simulation or malingering which has complicated the hysteria problem obtains a new significance in the light of this broader concept of the whole personality and the expression of the wish energy upon these varied pathways. It can readily be seen that the extension of a wish or purpose through the unconscious to all forms of action removes any line of demarcation as to where simulation begins or ends. Of course, there is such a thing as deliberate conscious intent to deceive and create the appearance of symptoms to attain a recognized personal goal. Or existing symptoms may be specially utilized or aggravated for such ends, but in such cases the distinction at once becomes obscured between such deliberate conscious intention and the unconscious purpose which is now believed to underlie all hysterical symptoms. Besides this point of view puts upon the physician a new obligation in regard even to the consciously practised malingering or simulation since that too must fall to a greater or less extent under the influence of the unconscious trends which make up the character and influence continually the conscious behavior and choice.

It goes without saying that there may exist a certain amount of the conscious simulation which if practised does not necessarily belong to the hysterical character and is utilized apart from any hysterical grouping of character traits and symptoms. Such superficial adoption of individual hysterical phenomena is, of course, comparatively easy of detection. This is particularly true because of the extension of the field of hysterical study which has put upon the physician the necessity of examining the hysteric in the light of the broader setting of both conscious and unconscious determination and all the manifestations which belong to such wider territory. Here also Freud's insistence upon the difference only in degree between the sick and the well makes of simula-

tion itself a subject of pathological study, not simply a question of one or another moral attitude.

STIGMATA.—The subject of stigmata has also dissolved itself in the more plastic modern conception. The idea of stigmata partakes of the demonology which once infused the subject of hysteria when patients bore certain marks of their possession which were universally recognized. Today, however, neurologists with a clearer understanding of the variety of symptoms and their cause in a fluid and changing energetic personality are at a loss to fix conclusively upon any symptoms or marks which can be considered distinctive of the hysterical individual. Janet made a distinction of anesthetics, amnesias, abulias, paralyses and character alteration as stigmata over against hysterical accidents under which he grouped hysterical attacks, somnambulism, unconscious or "subconscious" acts and fixed ideas, but these phenomena are too fluctuating in form and in degree to merit such distinctive definition. The factor of prime importance for descriptive purposes, more still for interpretation and for effective treatment, is the general reactive tendency which distinguishes the disease.

Such a type of reaction is possible in any person and the usefulness of such a type of reaction where repression, dissociation and conversion of dissociated material in a limited degree serve the purposes of daily life has already been mentioned. It would be an easy matter under special stress for any one to carry the use of these mechanisms too far, but ordinarily there is not sufficient occasion or stimulus to bring this to pass, or perhaps it may be better put, the accustomed conscious control and regulation according to the demands of a reality tend to resist such childish and ineffectual type of behavior. Thus it can be seen why some sudden calamity or undue strain or stress might act as the over-strong stimulus or undue strain or stress might weaken the resistance and allow the more impulsive form of action. Of course, this is particularly liable to take place in a character which has retained, unconsciously at least, more of its infantile emotional type of interest and reaction.

CHARACTER.—So psychically there is an unevolved character at the base of the hysterical reaction. Such a character has remained infantile or primitive in its thought and action. This means that it has not grown into that synthesis of interests and activities which direct all the tendencies and trends of a personality toward the seeking and attainment of adult goals with a chief delight in the strenuousness and incentive of a stimulating reality. The emotional life has not come under the control and guidance of the intellect to a recognition and appreciation of the real ends of life to be sought after and the sterner methods by which such reality may be attained. The childish occupation with emotional life through phantasy, largely unconscious, sets unreal values before the individual and presents a painful contrast between the phantasied attainments and those which actuality would necessitate. Hence a compromise must be made. This is not in the form of a healthy and workable adjustment to reality by which character and purpose are strengthened and reality is bent to effective individual attainment, but

is that partial psychoneurotic compromise by which the phantasied ends are indirectly and partially attained without too great offense to the conscious ideals of the individual and of society. Hysterical conversion is the result. The simplest illustration of this has come from the war when the soldier, his resistance weakened by fatigue, and finally confronted by the added stimulus of a frightfully difficult and sudden reality, found himself unable to "carry on." There may have been no real injury, but the personality, weary of the struggle which had been growing increasingly difficult to maintain the high conscious standard of honor and duty and to prevent the disturbing recognition of the natural but forbidden wish to escape the hard life, seized readily upon a motor pathway suggested by the temporary disability. Perhaps even an unconsciously remembered disability of some former time served to create a paralysis, a muteness, blindness, deafness which responded to no treatment that was not able to change the unconscious current of thought and purpose.

The suggestiveness spoken of is not that of mere persuasion or pithiatism. It is rather a biological readiness to follow some such path of least resistance or some pathway made familiar to the unconscious phantasy as soon as the external world seems also to present any encouragement to its adoption. Superficial persuasion may, of course, fall into the category of such external stimulus, opportunity or encouragement, but it forms only a small and unilluminating part of the deeper problem of biological readiness for such a type of reaction.

EMOTIVITY.—The instability of emotional manifestations is similarly explained. It too lies in the readiness of response in a wide possibility of sudden and ill-directed reaction which is present in such an infantile background which has not been brought into the synthetic and toughening control of an adult intellectual purpose. The personality has not experienced what Bergson calls the "growing organization of recollections with acts." It approaches that extreme type whom he theoretically describes as one "who lives in the past for the mere pleasure of living there, and in whom recollections emerge into the light of consciousness without any advantage for the present situation." Only the hysteric differs from this ideal dreamer in the fact that he finds, as Bergson suggests the dreamer would, such a life not workable. So the hysteric must suffer the pain of this realization and make therefore the compromise with his dreams.

It is this background of emotional readiness and the phantasies that prolifically multiply themselves to gratify it that form the soil for the suggestibility which has always been prominently emphasized in regard to the hysteric. Suggestibility is again merely a feature which is common to the mental life of all. It is due always to the affective ideas which lie ready largely in the unconscious where they are not yet taken up by a definite interest through the direction of the conscious thinking. These are susceptible to any invasion of interest, leading to thought or action, which may come from the outside world, particularly from another individual toward whom there is already an affective reaction.

such as confidence or some other manifestation of love. In the psychoneurotic this is often particularly a relation of dependence, an infantile form of love which makes for a peculiar susceptibility to suggestion, while as Janet has stated it the hysterical also are suggestible to certain types of action—those that tend to narrow the field of consciousness. This might be explained more clearly by admitting that they respond through their unconscious phantasy interests only to those suggested forms of action which they can draw into the unconscious purpose. Therefore their response does tend to withdraw them from the bringing together into the conscious purpose these interests into a synthesis of a different order. In the interests of reality and of the satisfaction of the cravings of the personality this should be one, tending always toward a wider outlook through consciousness.

The emotional reaction, which serves thus the unconscious rather than the purposes which relate to the external world, may manifest itself in a negativism, which may range all the way from a spirit of exaggeration, resignation and renunciation, on the part of the patient, to an antipathic refusal to eat, to converse, or to engage in any other expected activity.

EXHIBITIONISM.—A common method of carrying out the affective purposes of the unconscious is the hysterical endeavor to be always in the attention of others. Here the infantile exhibitionism stands in contrast to the adult type of candid self-appreciation which effects a place in society and strives to maintain it by efficient service both to self and society. The hysteric, feeling his or her own ineffectiveness, unconsciously magnifies infantile attitudes and methods to take the place of the adult endeavor. Slightest sensations are magnified to their utmost, somatic pains are worked to their limit and there is pathological attention to feelings and actions. This reaches at times sensational forms in startling accusations made against others, extravagant confabulations, self-mutilations and attempts at suicide, most often highly theatric. Some of these have almost the appearance of conscious attempts to deceive and produce an effect, but even in such case they are doubtless largely the work of the unconscious strivings which practice self-deception upon the patient even more effectually than they mislead the other members of society. While there is an apparent readiness to adopt and utilize as well as to manifest an apparently sympathetic interest in the common pains of humanity there is little real concern for these as they may afflict other people, but rather a reaction of jealousy and envy toward them. The negativistic phase of this self-seeking tendency, a masochistic martyrdom, is a frequent phenomenon.

MOTOR DISTURBANCES.—These are of wide range. The most striking of these are the convulsive attacks, and attention has always been centered upon these since the days when the same type of reaction in more primitive men and women set them apart as specially inspired by the deity. They have always been associated with times of religious fervor and have become epidemic under any occasion for great excitement. Certain individuals, perhaps more frequently women than men, may be

prone to such attacks, though this has been disputed. There is nothing new to add to the description of these attacks since that of long ago. They only receive now the more illuminating interpretation which makes them also part of the mechanism by which the unconscious ends are attained. Their frequency is relatively rare and in interest they tend to give place in modern neurology to the far more prevalent lesser motor attacks which have occupied much attention and demanded intensive study during the war. The convulsive attacks show in general a prodromal stage, one of muscular convulsions and a postconvulsive stage. The first stage shows the variety of manifestation to be expected in hysteria. It is generally speaking one of mental unrest with often some mental as well as motor retardation and a moodiness or feeling of distress. Then occur more special phenomena, localized tensions, palpitations, dizziness, auditory disturbances, all of which may occupy a longer or a shorter period before the convulsions begin. The attack may even end with these and never go on to the actual motor discharge.

The convulsions are extremely tonic in character, the tonus distinguishing the attack distinctly from the epileptic convulsion. The face assumes a dreamlike expression and the attitudes adopted often rehearse the phantasy content of the wish. The movements of extension and relaxation which pass over the body have always been interpreted as exhibiting a strong sexual element. In some the relaxation comes on quickly with many movements of contortion following and with great irregularity and variety otherwise of movement and action. The theatrical character of the convulsion may be manifested in a variety of individual positions and attitudes and activities, sometimes of a dreamlike and ecstatic character. Such have found place in religious manifestations. Facial movements may express much of this vast variety of phantasy interest which underlies the dramatic form of the hysterical exhibition and is therefore to be watched as indication of the meaning of this phantasy content.

Neurological examination after an attack reveals much that is distinctive. The pupils will be found widely dilated and usually react to light, although it may be found that this reaction is slow. The analgesia usually present makes sensory tests difficult. The skin and mucous membrane reflexes may be markedly diminished, but the deep reflexes show no marked alteration and signs of organic disturbance are absent. There is only rarely involuntary micturition or defecation.

The postconvulsive stage may be marked by a condition of lethargy which has given rise to the sensational tales of the "living dead." The lethargic state may continue only a few hours or longer, even for weeks. Functional processes go on in almost inappreciable measure, respiration being scarcely perceptible, the urine in some cases at least leaking away little by little with occasionally involuntary defecation, or these excretory processes may be suspended for some time. Other patients manifest a delirious dreamlike state, which may be of an active phantasy type. With some the dream states may be of a catatonic nature, with others of the manic-depressive type. The condition in which the patients waken

from their attack may be the starting point for much mental disturbance as to their surroundings, since the patient may have wandered away from home in the somnambulistic state or otherwise confused himself as to his environment, having only a very imperfect memory of what has taken place.

All this presents an example of the regression type of neurosis, for there is no effort here to establish oneself through the symptomatic pathway in social esteem or in biological potency, as Kempf has expressed it. Rather there is the effort to escape the struggle and to retire into gratification through phantasy production and utilization of these phantased forms of action, the dreamlike states. The regression in such manifests itself in its degree as belonging to an earlier stage of childhood phantasy gratification, or a still more infantile, even more archaic state as in the cataleptic conditions described. There may be dissociation phenomena as manifested in play of erotic phantasies revealed in the attitudes and form of activity, the postures and impulsive mannerisms and in the delirium, confusion and disorientation of the postconvulsive stage. The somatic conversion phenomena which belong to this regressive picture are shown in the lowered muscle tonus, anesthesia and the retardation, almost suppression, of the functional activities, such as respiration and excretion. Hence it can be seen that in the emotional reaction, as well as in the somatic phenomena, hysteria may range from the transformation of the slightly suppressed conflict which takes some other than the direct pathway to solution to the deeply regressive repression of the unconscious material which is then forced into deeply regressive pathways, ideational and emotional, and into deeply unconscious physicochemical manifestations.

In regard to the more distinctive conversion phenomena, the conversion of the emotional and ideational conflict into a variety of somatic symptoms, here again the various degrees of suppression or regression of the disturbing material may be represented with its reappearance in converted form at varying levels of the autonomic personality. The Charcot school with their later followers, to whom the war has given particular opportunity for these specific studies, have described very fully the various forms of tremors observable. The polymorphic form of the hysterical tremor is peculiarly diagnostic. Tremor may be simple and is frequently seen when the hands are at rest. Intention tremor is rare while static tremor is sometimes seen. The tremor is usually regular but may be instead irregular, ataxic, choreic-like. The localization is varied and when the muscles of the tongue or mouth are involved there results the characteristic stuttering or other speech disturbances of a pseudo-paralytic type. The tremor of paralysis agitans may be simulated by horizontal axis tremors of the head. Hand tremors which are frequent are of many forms. These tremors are usually exaggerated if conscious attention is directed toward them and they are aggravated by sudden emotional shock. They tend to disappear when the hand is in ordinary use as in eating.

Care must be taken in the presence of intention tremor, which is

sometimes present, to differentiate between it and the intention tremor of multiple sclerosis.

Other motor phenomena which manifest also much of the hysterical mental attitude are the hysterical hiccough, coughing, abdominal spasms, stuffing. There are affections of the respiratory organs which are more or less persistent, such as hysterical asthma, dyspnea, tachypnea, spasmodic aphonia, known also as laryngismus hystericus. There is also a classical hysterical shaking and a variety of lesser tics. Among all these motor manifestations can be seen all grades of the hysterical expression of the suppressed or repressed wish content and the conflict against it. Here too there may be a more or less evident conversion of the phantasy content into substitute activities, the source of which is barely out of consciousness, only suppressed from direct activity. Or there may be deep repression and a completely unconscious return of the dynamic wish through the unconscious reflex paths which activate the sensori-motor pathways of facial muscles or the vegetative reflex arcs which control respiration and other automatic functions. Only mental analysis can determine whether the disturbance lies only barely below the threshold of consciousness or falls under the repression neuroses. Without such investigation one runs the risk of removing by superficial means one expressed symptom only to make way for an effort on the part of the personality to react along some pathway which will be less obviously discovered and therefore less likely to be diverted from the unconscious aim, which is hostile to the conscious desire for health.

Analysis is an attempt to interpret the forms of manifestation through the different levels in the personality on which the conflict has chosen to express itself. It therefore gives both clearer understanding of the significance of the various forms of symptomatic expression and the depth and extent of their inroad upon the autonomic personality and must be the basis of therapy. For from this standpoint it is discovered by mental analysis what meaning lies behind the autonomic symptom which would express itself at any particular level of interest and of striving. Only thus can the personality be brought to a recognition of the futility of its chosen form of expression and be trained to bring the energy output up to an adult and real standard which can efficiently accomplish those purposes which make for the satisfaction and health of the entire, no longer dissociated, personality.

The same double aspect of more superficial, almost conscious expression, and of that deeper unconscious motivation of autonomic pathways is to be observed in such impulsive movements as those of hysteric crying and laughing, explosions of emotional irritability, as well as certain well coordinated activities like hopping, grimacing, etc. Care must be taken in these latter instances not to overlook a possible dementia præcox regression.

Besides the usually rhythmical movements so far described there are often irregular incoördinated movements which so closely resemble chorea as to deserve most careful diagnostic attention. Of course, there are sometimes combinations of hysteria with true chorea and the hys-

terical imitation of choreic movements has been well known. Rigid neurological examination however with attention to the reflexes, cerebellar and cerebrospinal symptoms, and the possibility of infection and fatigue or overgrowth should preclude mistake.

Large movements which involve whole related groups of muscles raise the question of a purely hysterical reaction or one which may be confused with a true myoclonus which is nonhysterical. At any rate this is another phenomenon which manifests the psychogenic appropriation of somatic pathways in a special form.

There is the same difficulty in separating a hysterical tetany from a true tetany. Investigation must be made into etiology, its relation to occupation, and Erb's, Trousseau's and Chvostek's signs as well as the calcium therapy test should be used, though the reflex tests are only of general clinical value. Electrical hyperexcitability of the muscles has been reported as a marked feature in the war hysterics which Babinski and Froment have separated off as disturbances of reflex order.

No reliable statistics can be given in regard to weakness or loss of muscular power in hysteria but this is one of the most frequent manifestations varying in degree and extent. Psychogenic, sensorimotor and vegetative paralyses must be definitely differentiated. Yet here again we are brought to the perplexing question that presented itself so frequently in the war neuroses and led to the setting off of Babinski's large group from the "hysterical" or pithiatric cases. Some attempt has been made to explain these upon some slight organic lesion, but these cases may be brought back to hysteria, if that deeper and closer connection between the organic and the psychic is kept in mind, possible through the stimulation of the reflex arcs, both the sensorimotor and the vegetative, by the psychic impulse, and which is in accord with the unconscious psychic desire. Here in many instances the disturbances are concerned only with mildly suppressed wishes and conflicts or these may tend to rouse into activity deeper, older impulses which then could utilize the still older pathways of reflex or autonomic activity.

Myasthenia in varying grades is present with most hysterics, which may signify the beginning of a paralysis or may simply remain as a weakness. While this may be mixed with a true neurasthenic myasthenia, the hysterical myasthenia is of purely psychogenic origin. It lies largely in the field of the mild suppressions and represents reaction to a feeling of insufficiency and inferiority, or fear of pain, injury, loss, or a reaction against the sense of unattainableness of desired objects, all of which exerts a depressing influence upon motor activities. Or it may more deeply represent the regressive type of inaction in the strongly repressed type of neurosis when the effect upon the motor pathways will be more profound. It is sometimes extremely difficult to separate such functional myasthenia from organic conditions. These are multiple sclerosis in its earlier stage, spinal cord lesions of an obscure location, obscure myelitis, tumors, certain toxic conditions, and a certain type of pressure neuritis.

The paralyses may appear in mixed form but the most distinct forms

are hemiplegia, paraplegia, and monoplegia. All these have been particularly prominent through the war neuroses, showing how ready they are to serve as conversion phenomena for the mildly suppressed conflicts occasioned by war conditions.

Hemiplegia is present in about half of the cases of the total paralyses, according to Ziehen. It occurs more often in an incomplete form, diplegic, in the arm and leg but without cranial nerve involvement, or rarely the cranial nerves may be involved. The presence of the knee jerk and the Achilles jerk, absence of atrophy, hypotonus, trophic disturbances, etc., testify to the non-involvement of the peripheral nerves. There may be a mixed hemiplegia involving the opposite arm or leg. Very rarely are there alternate hemiplegias or quadriplegias. The hemiplegia is almost invariably ushered in by an affective shock; sometimes it develops after a major hysterical attack. Usually a sense of giddiness and weakness is associated with the onset and sometimes pain, nausea and vomiting are present.

Experience proves that too great care cannot be exercised in determining a hysterical paralysis or an organic condition. Tendon reflexes should remain unaltered, clonus, increased tonus and other signs should be absent, showing that the cerebral neuron is not affected. Yet the knee jerk, according to Babinski, is usually very marked and ankle clonus, called by some authors pseudo-clonus, has been reported present. Other signs too once considered decisive of organic lesion are found to be present, such as the presence of inferior cutaneous reflexes, foot tremor, foot clonus. All of which points to the possibility of a closer similarity in the result of functional interference because of actual lesion and that effected by the psychic utilization or misappropriation of the autonomic paths by the unconscious impulses.

Although the motor cranial nerves are rarely involved, cases are reported of oculomotor and abducens paralysis. It is possible that here the difficulty of distinguishing between a paresis and a spasm has caused a mistaken diagnosis. Cases of ophthalmoplegia externa should be considered in the light of hysterical contractures. Hysterical ptosis is fairly common.

Facial palsy is a special hysterical symptom. All branches of the facial nerve may be involved though less frequently the supra-orbital portions.

The glossolabial involvement may show mixed spasm and paresis. The movements of the palate are restricted in observed hysterical aphonias. Care must be exercised in the appearance of inferior alternate paralyses with crossed facial.

Paralysis of the larynx is usually incomplete though it is more often bilateral than unilateral.

Paraplegia manifests itself most frequently in the lower limbs. There may be complete or partial inability to move the limbs or an inability to move them in the vertical position while they can be moved in the horizontal position. These paraplegias may be of the flaccid or spasmodic types and contractures are often present. Distribution may be extensive

and yet restricted movements be preserved. Babinski and Froment have described the striking appearance of these phenomena in their war cases. Indeed trauma is a very frequent cause of such symptoms, even among children, perhaps on a basis of fatigue of the parts involved. There is often here exaggeration of the patellar reflex. One must be on guard here against confusing a condition due to minute hemorrhages in the spinal cord.

A particular form of paraplegia is manifest in the inability to walk or stand, though no paralysis or ataxia is manifest in the sitting or reclining positions. This may be called a psychogenic astasia-abasia.

The very frequent hysterical monoplegias are very irregularly distributed. Perhaps brachial monoplegias are the most frequent while those of the legs are rarer. There may be affection of the cranial motor nerves as mentioned, or of individual muscles innervated by a nerve group. There may be then various ptoses with perhaps nystagmus.

SENSORY SYMPTOMS.—A hysterical pseudo-tabes presents some difficulties, especially as hysterical symptoms may be grafted upon a true tabes. Purely hysterical may be pains, incoördinated ataxic movements, analgesia, eye disturbances and other symptoms. The diagnosis, however, can usually be correctly made.

The question of anesthesia is a disputed one. The effect of the psychogenic origin of the disturbances is much in evidence in the variability both as to location and to degree of the anesthesia. Incomplete anesthesia or hypesthesia is more frequent than complete loss of sensation. The reports of anesthesia have been much exaggerated through the difficulty of obtaining reliable testimony from the patients and the influence of the dream state conditions.

Total general anesthesia is probably very rare. Anesthesia may be unilateral or it occurs frequently in patches in isolated areas or in symmetrical regional areas. Hemi-anesthesia or hemi-hypesthesia frequently exists and the left side is the more frequently affected. Affection of geometrically limited areas, rarely coinciding with anatomical nerve distribution, is well known. Crossed anesthesia may exist but care must be taken to distinguish this from the result of possible minute thalamic lesions. The anesthesia is little noticed by the patient as a rule although occasionally there may be pain or a pricking or a crawling sensation or a feeling of numbness as if the limb had gone to sleep. Anesthesia is usually of a sudden origin and offers much room for suggestion as an instigating factor.

Closely associated with disturbance of tactile sense is the loss of ability to distinguish heat and cold, though the hysteric always responds strikingly to cold application, particularly in a shower or by packs.

Deep sensibility is only rarely disturbed. Occasionally there are dissociated conditions with loss of one sensibility, tactile or deep, and retention of the other, but this is rare.

Disturbance of the mucous membrane sensibility is similar in manifestation to that of the skin.

Pain sensibility may be diminished or exaggerated. Absolute anal-

gesia occurs only in an occasional ecstatic dream state. Hemi-analgesia and hemi-hypalgesia occur very much as hemi-anesthesia and hemi-hypesthesia with also crossed varieties. The intact pupillary response to pain stimulus testifies to the unimpairment of the reflex mechanism even if the pain is not recognized by the patient.

Hyperalgesia is most common. Certain classical points of pressure exhibit marked tenderness and have given rise to much discussion of so-called hysterogenic zones. These, however, seem to emphasize the psychogenic nature of the symptoms and their relation to emotional stimulus as they concern areas in which special physiologic stimuli are set up with a special readiness. Pressure on the same points or others may cause the attack to cease or the localized pains, paralyses, contractures, or what not, which have been roused, to disappear.

Localized and spontaneous pains show also a great variability but are of very frequent occurrence. In the topalgias there is abundant testimony to the reflex visceral disturbance brought about by psychic factors. Of course, many of those considered as hysterical are coming to be more clearly recognized as due to actual visceral disorder acting upon the reflexes.

The neuralgias particularly have received much more careful recognition recently as due to organic causes, such as cervical rib or other anatomical anomaly or diseased condition such as diabetes.

The ready effect of suggestion therapy would seem to testify to the existence of hysterical arthralgias, but diagnosis of these has been much restricted since the radiograph has been used in coxalgias and other forms of arthralgia.

The existence of hysterical enteralgia is supported by the frequency of hysterical vomiting and several forms of severe visceral pains are of undoubted psychogenesis. Simulation has been called in to explain hysterical ileus with pain and fecal vomiting, but here again there is a true hysterical condition which is due to the influence of unconscious rather than conscious factors. The psychic factor probably also plays an important part in hyperemesis gravidarum. Archaic and infantile confusion in phantasy of the reproductive and nutritive pathways plays a large rôle in the gastric and other alimentary disturbances.

Hysterical cardalgia is very frequent and from the standpoint of unconscious disturbance of functional pathways it can readily be seen that this would be the case. It may occur in a severe form, that of angina pectoris hysterica, or in a variety of lesser symptoms, pains, discomfort; interference with heart action as evidenced by the pulse, etc. This belongs among the vast group of gastric, enteric, genital, and other false pathies which are easily to be seen as under psychic control. For these functional pathways are not only at psychic disposal but are also largely concerned in group fashion with the functions of the personality. There may be also a connection here with actual neurasthenic conditions.

Not enough is known as yet of organic disturbances of the vestibular apparatus to admit of a sweeping inclusion of apparently hysterical manifestations under the hysterical diagnosis. There are no doubt hysterical

manifestations associated with some other cause, gastro-enteric, fatigue of eyes or other parts, etc. There is the possibility of a hysterical reaction due to cerebellar vestibular disturbance.

Disturbances of smell and taste have not been satisfactorily investigated. Ageusia may accompany facial anesthesia. In this field there is also obviously room for wide variation under control of the psychogenic factors.

Hysterical deafness and deaf mutism have been apparent in the war hysterics but these symptoms are usually comparatively rare. Yet bone conduction is found to be modified and certain investigators have reported absence of modulation of the voice which is characteristic of deafness. Here, however, there is ample room in voice production for distinct affective alteration of ordinary modulation in accord with unconscious impulses and trends.

Unconscious influence upon sight has been strikingly illustrated in a few instances and in general tests have revealed the somatic unimpairment of sight and proved that it is only psychically affected. Unilateral blindness is the more frequent, although there have been a few cases of double-sided blindness. Diminution of vision, which usually affects the left eye, appears to be a purely psychogenic affection and largely due to suggestive examination. It is a complaint that arises largely out of fatigue or an inability to concentrate attention.

Scotomata and disturbances in color perception, pupillary immobility and unequal pupillary reactions are sometimes observed among other complaints which the hysterical reaction is able to produce.

VEGETATIVE SYSTEM CHANGES.—In the field of vasomotor and trophic as well as secretory disturbances there is still much discussion. Here is opportunity for closer investigation of the relation of the activating wish stimulus and the autonomic pathways over which these operate.

Thermic changes show great variation and an anomalous character. There may be rise of temperature or there may be localized hypothermia, which was observed in Babinski and Froment's cases of so-called reflex disturbances. Skin changes, urticaria, edema, and other secretory disturbances are all to be considered carefully as manifesting a purely psychogenic origin or a psychic activity upon a constitutional condition of skin irritability, trophic peculiarities or what not. Here again the particular psychic trends of the personality may utilize the physiologic pathways and at the autonomic level of nervous activity convert an auto-erotic tendency, an exhibitionistic wish, any primitive self-seeking type of reaction into such definite disturbances far below the level of conscious choice or of conscious awareness. Occasionally even the hysterical aggravates such conditions or induces some such manifestation, as a skin disturbance, by an interference and activity which is almost clear in consciousness as an impulse or an attitude, so that these pathways of manifestation are at the disposal both of the suppressed and the repressed factors of the personality.

This may be true also in regard to any secretory activity. There may be increase or diminution of saliva, sweat, of the flow of tears, while

aggravated bronchial and nasal secretion is a most common symptom. All of these are indicative also in the healthy of the ready conversion of emotion when it is roused on certain occasions. It must, however, be kept in mind that it is possible in these manifestations to have a disturbance due to multiple sclerosis or syringomyelia.

Both the urinary and intestinal secretions may be much modified in hysteria and form also ready avenues, associated as they are with infantile interest, and also with the adult sexual, for the unconscious manifestation of such interests which under repression seek some related conversion pathways.

Changes in metabolism also fall under the same form of explanation as results of the disturbance of function by the psychic pressure which has all somatic pathways at its disposal.

Alterations in reflexes are of but little diagnostic importance, though they are frequently present. The corneal, conjunctival, palpebral reflexes are diminished and there may be loss of the faucial and pharyngeal reflexes. The triceps and radial periosteal reflexes usually are increased. The anal reflex may be absent or show hyperesthetic sensibility, like the rarer vaginal reflex. These two are evident signs of the emotional disturbance of the reflexes. Marked irregularity in the reflexes should suggest organic lesions.

The knee jerk and Achilles jerk may be lost during or after hysterical attacks, though they are as a rule increased in hysteria. Here, too, the method of interpretation is not yet sufficiently clear.

PSYCHOTIC SYMPTOMS.—The strictly mental manifestations have been freely mentioned throughout this discussion in their close association with the various forms of conversion phenomena. Yet special attention may be given to certain mental states which may mark the hysterical picture and which show more or less profound mental alteration as a result of the unconscious striving. The so-called hysterical dream state or twilight state has become well recognized. Here consciousness has fallen temporarily to such an extent under the control of the unconscious phantasies that the patient remains or goes about in a state of dreamy delirium or in a deep revery, retaining a general orientation or an ability automatically to engage in ordinary conduct but with complete or partial amnesia afterward for his acts or for the condition. The dreamy attacks may be associated with a major seizure, immediately preceding the latter or existing during the attack. In this case the patient may be quiet and self-contained but dazed, or there may be the profound lethargic condition already commented upon. In the automatic activity of the state of revery patients may go far from home and be temporarily or permanently lost from their surroundings, or they may commit crime, and even criminal acts which have a motivation in consciousness may be carried over to commitment in this dream state, so that this state is a most important matter of medico-legal interest. Pathological lying, accusations, and the peculiar spasmodic, impulsive type of talking, laughing, emitting animal cries, etc., which has been called by

Ganser "Vorbeireden" belong also in this category of twilight state acts.

Care must be exercised to distinguish this state from the similar one belonging to alcoholism, epilepsy or traumatism. The dreaminess associated with an appearance of consciousness, together with the suggestibility, rapid changeableness of conduct, the foolish, childish character of such conduct and the signs of somatic conversion distinguish the hysterical dreamy state.

The presentation of such a variety of symptoms and also the ready ability to adopt symptoms from any other diagnostic group offer special danger of confusion with other psychoses.

It is becoming more and more clear under penetrating research that there is not the close resemblance in the convulsive seizures to epilepsy once believed. Of course the two syndrome groups may be present at the same time in the individual. Aside from the distinctions which may be observed in the seizures, in which the loss of consciousness and of motor control is complete in epilepsy and partial and incomplete in hysteria, the attempt to separate the two diseases in the light of the dynamic trend makes a clearer differentiation between them. At the same time it gives the affective background out of which the impulses arise as a similar one, utilizing however different mechanisms for taking up the dynamic impulse. Epilepsy belongs to the regressive type of neurosis, perhaps appearing in a hallucinatory, dissociated form of phantasy, but at any rate representing a drift back to an earlier biological level, psychic and somatic. This manifests itself in the completeness of the attack in its return of the patient to an infantile or preinfantile level and in the gradual permanent regression of psychic and physiologic function as the epileptic deterioration proceeds.

Hysterical features may appear in connection with any other psychosis, since hysteria manifests a type of reaction in an altered personality. Somatic organic tests are a criterion for a differential diagnosis. In the more evident mental reactions more difficulty is experienced but here too the consideration of the differing mechanisms is a guide. Hysteria is close to dementia praecox in its manifestations, yet as Jung has shown, these two syndromes represent different direction of reaction. In the hysterical there is still the tendency to activity, to reach reality although by the pathways of displacements and conversions, while in the dementia praecox the pathways are blocked and the trend is away from the real world. Or according to Kempf's interpretation of the same distinction, in dementia praecox the patient is unaware that it is the ego's own wish and craving which cause the distress, that the latter has a personal source, while the hysteric is able to acquire this knowledge. Kempf includes the latter in the "benign" type of neurosis and the former in the "pernicious" type.

Treatment.—The treatment and the prophylaxis of hysteria are most important considerations. The history of hysterical therapy would have to begin with the history of the disease itself, which we have seen is practically simultaneous with that of the human race. Just as the

conception of hysterical manifestations has passed through many stages, so has the practical approach to it. Its dynamic nature, as that is understood today, explains also the method of sensational, "miraculous" cure which has not yet passed out of possibility. Anything that will slowly or suddenly alter the form of the striving for the active cravings and through confidence, "*faith*," open the personality into outgoing channels such as a trusted or loved object offers to every human being can make an occasion for a cure. This is in essence the "transference" phenomenon of Freud. Such a redirection of energies gives at least a partial and often a most striking relief of affective cravings, by which healthy function has been restored.

Suggestion and hypnosis have the same basis for their effect. They too represent the readiness of the unconscious self to expand, to open up to outside objects through the stimulus of emotional response to the physician or other medium of healing, and an alteration takes place both in the goal and the mechanism for reaching that goal. The change in all these cases is really in the patient himself and effected only by him, although by his unconscious self where after all his dynamism lies. Thus suggestion, hypnotism, the presence of a "divine" person or a sacred spot, or the sudden event tragic or otherwise which changes the trend of the striving, are all only opportunities for this personality to assert itself in new ways, ways which better fit the demands of the real world upon the personality.

There exists a danger in all these methods of only partial alteration of the personality. A seeming cure may be effected in regard to a portion of the personality, while other trends, other cravings still conceal themselves. Some childish pathway is still overcharged and over it the wish bursts forth later in a new symptom or group of symptoms.

Freud recognized this together with the need to reach the more persistent symptoms which were not accessible to the methods mentioned. He therefore instituted and developed the slower but more thorough method of *psychoanalysis*. He bases it upon the same principles which underlie the forms of therapy discussed, only he carries it through the analysis to the point where the aims and strivings of the personal cravings are brought to the clear knowledge of consciousness, so that the patient has a conscious control and choice of methods of reaction before him and is no longer unconsciously driven to utilize the ineffective and distressing ones of the hysterical mechanisms. Also the patient is left with such a wholesome knowledge of the mechanisms once employed and the manner in which they are built up and employed by the unconscious that he can avoid their domination and so control his own life without the need of such indirect "symptoms."

Freud's chief medium for bringing the patient to a knowledge of the hidden affective cravings as well as their manner of transformation over into substitutions and somatic symptoms is the dream. He examines this in the light of the free associations with which the patient surrounds it in the conscious telling and contemplation of it. In this way a knowledge is gained of the whole personal aim and trend with

these individual factors which have been pathologically developed. It is out of this background of understanding and approach that Kempf has developed his classification in which he aims to make more clear the dynamic trends which the autonomic affective cravings of the personality have taken. In this way not only can the mechanisms be understood, but they can be followed out therapeutically with the patient. Thus other more effective mechanisms can be substituted and the craving or wish brought again into a form of activity which satisfies both the personality and its relation to its social environment. This, of course, is the process of sublimation and removes the barrier erected by the faulty mechanism and through the phantasy form of the wish. It finds instead an adult form of direct gratification in many forms instead of the impossible and unacceptable one which was harbored in the infantile phantasy.

This sort of classification gives also the best understanding of the prophylactic possibilities in regard to hysteria. Freud has made it plain that these lie chiefly in the period of childhood, and consist, most simply expressed, in the aiding of the child to develop in a free and unhindered attitude toward the world of reality in which he must live. This is not the unguided and untrammelled following of individual impulse, for this would only increase the introversion upon the interests of self and the intensifying of these interests to the exclusion of relation to other selves or the outside world. Then when the child in its earlier or later years finds that these interests must be altered and adapted to meet external conditions it is driven away from the difficulties which the latter present and back into an unconscious, in the hysteric a partly conscious, indulgence in phantasy gratification instead of the efforts and often hardships of reality. Then the struggle continues between the cravings and the phantasy form they have taken and the conscious world so that the conversion or substitution pathways are resorted to.

It is therefore important that children should be treated with that wholesome neglect which does not set them in the limelight of importance from which it is so hard for the immature adult later to free himself and which makes the hard contrast between the ideals of phantasy and the actualities of the world of environment. On the other hand a too great actual neglect may foster in the child a sense of deprivation and abuse, a negative or masochistic sense of self-importance also productive of phantasy exercise rather than reality. In either instance account must be taken of the inherited constitution including possible inferior physical constitution and of other handicaps which make the adaptation to the external world more difficult and phantasy production easier.

Most important is it to watch the attitudes which the child builds up to definite questions and situations of life. The child is confronted with facts and situations which in the light of his inexperience, ignorance and comparative helplessness present to him far more serious and weighty problems than adults usually remember as belonging to their own history.

Chief among these or forming the center of all his problems is the

sex problem. Here again Freud has not only brought to attention what children themselves clearly enough reveal to us when we have eyes to see it or courage to believe it, that the sex problem is always with the child and of paramount interest. Moreover Freud has taught us to recognize it not in a simple unified adult form but in a polymorphous form in the many appeals it makes through the child's own variety of interests in its own body, and in its difficult relations of adjustment to the objects and individuals, chiefly the parents, by whom it is surrounded, and its love response to them. It is a variable problem also because the child is passing through various stages when one form of sex interest or another is paramount on its way up to adult maturity. Here more than anywhere the judicious but candid presentation and treatment of facts aid the child to develop in the realm of reality rather than phantasy and to have a wholesome attitude of handling those facts and building up reactions which are free and self-expressive and do not necessitate the roundabout pathological mechanisms of substitution, conversion, etc.

The prophylaxis of hysteria chiefly concerns childhood since the reactions are deeply laid and for the most part dependent on a lifelong faulty attitude. Of course, for a hysterical nature, one in which such an attitude is present, there can sometimes be avoidance in later life of occasions of special difficulty which put more upon the individual than he can bear and so loosen the hysterical reaction. Here, however, it must be considered that such protection of the hysteric may be only the gratification of the unconscious weakness and dependence, phantasy gratification for the individual. This should never take the place of a therapeutic effort to discover and remedy the inner features of the difficulty. In the case of a mild suppression hysteria, such for example as war conditions produced, this might be avoided by avoiding conditions of extreme hardship and fatigue without which the individual might always maintain a healthy reaction. In many cases, however, there would be found an already imperfect mechanism brought more prominently into activity by the difficulties encountered and the temporary personal debility.

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CHAPTER XVIII

HEADACHE

By E. BATES BLOCK, M.D.

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Definition.—*Headache*, known also as *cephalalgia* and *cephalea*, is a symptom of many diseases, and consists of an aching sensation in or on the head.

Etiology.—*Heredity* plays an important part in headache. Campbell thinks this due not only to the inheritance of headaches but to the factors which cause the headache, such as "ocular, nasal, and dental defects, uric acid, nervous and other diatheses."

Age.—Functional headaches are very uncommon before teething begins but may occur during teething, whether in the first or second dentition, or in cutting the wisdom teeth, and is probably due to fever. Headache is frequent from four or five years upward. Treichler says one-third of the schoolchildren in France and Germany suffer from headache (Campbell). Puberty is a very common time for headaches, migraine, chorea and other neuroses to develop.

Headache says a continuous headache may occur soon after the skull closes, and will gradually subside.

Sex.—According to Campbell, during the reproductive period headaches are more common in women than in men. This has been estimated at 76 to 16, and again at 65 to 26; migraine is said to be slightly more common in females than in males.

Season.—Winter gives most trouble because of exposure to colds, draughts, etc.

Weakness may produce headaches, as after prolonged illness, hunger, abstinence from coffee to which one has become accustomed, especially in the morning. Over-work probably produces headaches from cerebral congestion. Excessive exercise, coughing, blowing on wind instruments, and excessive laughter or crying, and fatigue may cause headaches, and render a preëxisting headache worse. Emotional causes, such as anger, worry, excitement, are not uncommon.

Defective Ventilation.—Sleeping in a closed room, or living in crowded, poorly ventilated rooms, is a frequent cause of headache. Espe-

cially is this true when the room is heated by a stove. Excessive moisture in the atmosphere is said to provoke headaches, but the author has seen headaches often from excessive dryness from steam-heat radiators. A low atmospheric pressure is said to cause frontal headache.

Time of Day.—Headache is often worst in the morning. The patient wakes with it. This has been attributed to the uric-acidemia of the early morning (Haig); poor ventilation; mouth breathing; the horizontal posture (Campbell). But it is more often due to (1) the prolonged period without ingesting fluids or food, and the lack of dilution of poisons in the body; (2) a restless night; (3) not having the neck properly supported on a pillow. The middle of the day is the most comfortable time for people with headaches. Headaches from eye-strain are very apt to come on about eleven or twelve o'clock in the day, although the patient may wake with them. Headaches from nasal causes are usually present on waking in the morning. Headaches occurring in the afternoon and evening are often syphilitic, or due to fever rising at this time of day, or to over-work and fatigue. Rheumatic headaches are said to be worse at night.

Temperature.—An over-heated room may cause headache. Insolation may cause most intense and lasting headaches, after which the least exposure to the sun or fatigue causes violent headache. These headaches may often last many years; in one of the author's cases it was associated with epileptiform convulsions. Exposure to cold or "sleeping cold" often causes headache. A very evanescent pain in the frontal region is sometimes experienced from eating ice cream, or taking very cold drinks. "The atmospheric state preceding or accompanying a thunderstorm is a well-known cause of headache" (Campbell). "Any sharp, biting wind is apt to cause it." In neuralgic headaches the east wind seems to aggravate most.

Organic Brain Diseases.—Headache is a common symptom of meningitis, encephalitis, encephalomalacia, brain tumor, brain abscess, chronic hydrocephalus, oxycephaly, osteitis deformans, general paralysis of the insane or functional brain diseases, such as hysteria, neurasthenia, hypochondria, or after epileptic attacks.

In brain tumor the pain is produced (according to Byron Bramwell) by (a) increase of intracranial tension and consequent stretching of the membranes; (b) direct involvement of the membranes, periosteum or bones; (c) direct implication of the fifth nerve. Meningitis produces most agonizing headache, and may be followed by persistent headache, which may last a lifetime. In acromegaly the headache is usually frontal. When it occurs in cerebral hemorrhage, it indicates a meningeal seat. It may occur also in cerebral thrombosis.

Injuries to the head, such as a jolt or a jar or direct trauma to the head, may cause headache.

Fever.—Headache is often the first evidence of fever. It may occur in fever from any cause. Not only the height of the fever but also the kind of disease producing it influences the occurrence and severity of the headache. Sometimes the acute infectious diseases, such as typhoid

fever, pneumonia, influenza, etc., are followed by persistent headache. It is not improbable that some of these cases represent actual meningeal involvement which may be mistaken for the delirium from fever. Malaria may be followed by persistent headache, probably due to anemia, or headache may occur from the fever during the paroxysm.

Sleep.—Insufficient or interrupted sleep may cause headaches, or sleeping with the head in an unnatural or strained posture without proper support of the neck. In some people a mid-day nap or over-sleeping will cause it.

Lithemia or gout is a common cause of headache and is often associated with dizziness.

Anemic headaches, from general anemia, are well known. Severe hemorrhage, as from injuries, childbirth, etc., may produce severe general headache. The headache is usually frontal, but it may be in the vertex, general, temporal, or occipital. While the headache is more often on both sides, it may be on one side only, more commonly on the left side. The headache of anemia from prolonged lactation, leukorrhea, poverty (inanition), chronic suppurations, etc., is well known. The headache is often continuous, with some neuralgic pains and throbbing. It is not due to the small quantity of the blood, but to its poor quality. It is usually helped by lying down.

Headaches from hemorrhage, chlorosis, and other anemic conditions are usually dull and heavy, seldom severe. Headache occurs also in pernicious anemia and secondary anemia.

Intestinal parasites probably cause headache from anemia.

Active Hyperemia.—Colin suggested as causes: too thick or too tight hair; violent bodily exercises, as running, laughing, coughing, vomiting, defecation; violent emotions, such as intense anger, grief or shame; and excessive intellectual toil (Campbell). The headache is often associated with throbbing, vertigo, tinnitus and flushing of the face. Active congestion, such as occurs from violent emotions and other causes, may aggravate an existing headache or excite it in those predisposed to it.

Passive Hyperemia.—This may be produced by diseases of the heart or lungs, muscular efforts of the trunk, or constrictions around the neck, but Campbell thinks some other factor also necessary to produce the pain.

Headache from hyperemia is made worse by bending over, coughing, straining, sneezing, blowing the nose, etc.

Posture.—When there is congestion of the head during headache, stooping over or lying down makes it worse; any sudden change of posture may aggravate it temporarily, but as a rule lying quiet ultimately affords more comfort and relief than being up.

The Scalp.—Pediculosis capitis, which is associated with enlargement of the cervical and occipital lymph-glands and the glands back of the ears, and probably indicates interference with the lymphatic circulation, may cause headache. The occiput and vertex are especially affected. Pulling the hair too tight in arranging it is a common cause of headache among women. The weight of the hair is also suggested by Camp

bell, or wearing a heavy hat. Osler is credited with saying that headache has been attributed to "everything from an adherent clitoris to a high hat."

Earache is a common cause in young children. Loud noises, especially artillery practice or large gatherings, may cause headaches, and always aggravate them. Any disease of the ear, especially inflammations, may give rise to headaches. The pain may radiate to the whole side of the head, to the temple, parietal region or occiput, or it may even extend over to the opposite side of the head. Obstruction of the eustachian tube causes headache by reduction of the intratympanic pressure. Foreign bodies in the ear, or wax, occasionally cause headache.

Teeth.—Apical abscesses, pyorrhœa alveolaris, dental caries, or impacted teeth are said to cause headache, although the tooth itself may not be painful. The upper teeth are more liable to produce headache than the lower, especially the molars. Headache from the teeth may be frontal, temporal, supra-orbital, or orbital. The lower wisdom tooth usually produces pain in the ear or temporomaxillary joint. The pain from impacted or carious teeth or abscesses may be occipital or nuchal. Dental headache is more often unilateral than bilateral.

Eyes.—The eyes are probably the most widely known cause of headache. Bright light, such as sunshine, snow, or the glare on the water, may affect the optic nerve quite independently of the reflex effect upon the intrinsic ocular muscles, according to Campbell. The visual system is peculiarly sensitive in many diseases of the eye, such as conjunctivitis, keratitis and iritis. Various organic diseases of the eyes, such as glaucoma and tumors of the orbit, cause headaches; the pain is usually only on the side of the eye affected. Any increase in intra-ocular tension will cause headache. However, atropin increases the tension but sometimes relieves headache, due to lessening the strain. Eserin diminishes the tension, and may afford relief to headache. Eye-strain is due to the act of accommodation, focussing and fixation. Headaches are usually produced by fatigue of the intrinsic or extrinsic ocular muscles. Strain of the intrinsic muscles causes frontal headache, while strain of the extrinsic muscles causes occipital headache.

Of the refractive errors, hypermetropia is the commonest cause of eye-strain (more common than myopia or astigmatism). Hypermetropia and astigmatism throw more work on the ciliary muscle. Myopia causes headache probably in part from over-work of the adductors. Weakness of the ciliary muscle may be due to paralysis, presbyopia or loss of tone from general debility, anemia, neurasthenia or any debilitating disease. In debilitated states of the nervous system the headache is usually vertical, but if brought about through the eyes may be frontal. Asthenopia may be due to food poisoning, and may be improved or corrected by diet or attention to the bowels; it is said by Campbell to be due sometimes to nasal disease. Paralysis of the extrinsic ocular muscles often does not give rise to any headache because fixation is impossible and is therefore not attempted, thereby occasioning no eye-strain.

It is particularly in the milder forms of muscular weakness that headaches occur, due to the effort made in fixation.

According to Crawford and others, hyperopic errors are more common in children than emmetropic and myopic conditions.

Walton questioned 100 individuals with healthy sight (of whom 31 per cent. never had had headaches); 42 cases of partial blindness acquired or congenital (of whom 29 per cent. never had had headaches); and 90 cases totally blind since infancy (of whom 66 per cent. never had had headaches). Headaches, therefore, seem twice as common among those subject to eye-strain than in those in whom this element is wanting.

According to Wilson, disturbance of muscle balance was present in 60 per cent. of his 200 cases of headache, and the prevalent variety suggests over-taxed or feebly developed accommodation.

In 200 cases of headache due to ocular disturbances, Wilson found: Age, 10 to 49 years; sex, 177 females to 23 males; occupations, over one-half were dressmakers, typists, scholars, clerks or students; locality of pain, order of frequency was: forehead, behind the eyes, vertex, occiput; time, some had it all day, in some it began in the early morning, and in many it was associated with near work only; vision, 68 per cent. had good or fair vision, 19 per cent. moderately bad vision, 13 per cent. had bad vision.

Refraction:

Emmetropia	20	per cent.
Low hypermetropia	24	" "
Low myopia	9	" "
Hypermetropic astigmatism	20	" "
Hypermetropia	11	" "
Myopic astigmatism	7	" "
Mixed astigmatism	3	" "
Myopia	3	" "
Others	3	" "

Most of the cases of emmetropia had frontal headache. Heterophoria existed in 60 per cent. of the cases, of which esophoria was the commonest (37 per cent.).

Muscle Balance.—Doyne says that pain at the nape of the neck (nuchalgia) is perhaps always due to disturbance of muscle balance (probably meaning, if due to eye-strain at all). Wilson found that nearly all of his cases who complained of pain at the back of the head or neck had disturbance of muscle balance. Doyne thinks that the exhaustion of the automatic power of the eyes for fine adjustment in anisometropia is the main cause of headache. Wilson does not think there is much evidence of preference as to what form of eye-strain produces headache. Anisometropia occurs chiefly in hypermetropic astigmatism, myopic astigmatism, hypermetropia and mixed astigmatism.

Diseases of Nasal and Accessory Sinuses.—Skillern says the cause of headache in nasal and accessory sinus troubles depends upon "(a)

swelling of the mucosa with pressure or irritation of the nerves; (b) direct contact of the swollen mucosa; (c) negative pressure in the sinus; (d) stasis following obstruction of the drainage passages; (e) ulceration of the mucosa with involvement of the nerves; (f) reabsorption of toxins formed within the sinus; (g) any condition which causes active congestion of the cranial circulation (acute exacerbation of a chronic inflammation, over-indulgence in alcohol and tobacco, etc.); (h) disturbances in the blood and lymph circulation at the base of the skull."

It may be brought about by acute coryza, nasal catarrh, nasal calculi, and even worms are said to get into the nose and frontal sinuses and cause headache. Morgagni, in commenting on Bonetus Sepulchretum, says: "And with God's leave even scorpions" were found within the skull, which he did not believe. (Quoted by Campbell.)

Diseases of the frontal, ethmoidal, maxillary, sphenoidal and tympanic sinuses, such as polypi, foreign bodies, and inflammations often cause headache. These may persist for years without a diagnosis being made. Probably the most common are nasal polypi, and swelling of the turbinate bones, very frequent with hay-fever.

Autogenous Poisons.—Constipation is one of the commonest causes of headache and is usually thought of by the laity and physician before anything else. Many people never have a headache except from constipation and always have it when the bowels fail to move. Daily movements are no certain indication of bowel health, as the movement may be twenty-four hours too late. Ileocecal insufficiency is very important.

The Kidneys.—Uremia often causes headache; it is said to occur most often in the morning. Headache may precede the uremic attack. In some cases they are due to meningeal edema. Renal calculus sometimes causes headache. Headache occurred in 36 per cent. of the author's cases of Bright's disease, as seen clinically. In all cases of headache the urine should be examined, and especially the degree of acidity, acetone, diacetic acid, indican, urobilin and urobilinogen and oxaluria noted.

The Liver.—Biliousness, jaundice, and acute yellow atrophy are causes of headache.

Uric acidemia is often a cause of headache and migraine; the headaches are apt to occur during the morning alkaline tide.

During headache the *stomach* often ceases to absorb; any food or medicine in it will lie there until vomited or until the headache passes off. This fact has often given rise to the idea that the stomach causes the headache, which is not usually true. No doubt exists, however, that certain foods will produce headaches in certain people, especially when they suffer from indigestion; for the food spoils in the stomach and the poisons that are thereby absorbed produce the headache. It is particularly in cases of achylia gastrica that this occurs. In cases of hyperchlorhydria associated with headaches, they are both probably due to the same nervous cause. Dilatation, gastroptosis and fermentation are often associated with headaches.

Acute and chronic gastritis, and intestinal indigestion and intestinal

worms may cause headache. Stricture of the rectum is said to cause pain in the occiput.

The headaches in *diabetes* are particularly severe and persistent. Among the glands of internal secretion, headaches occur in acromegaly, myxedema and Addison's disease.

Exogenous Poisons.—Headaches are often due to *exogenous poisons*, such, for example, as nitroglycerin, which produces a feeling of fullness in the head, and either pressure or a lifting sensation in the vertex. Quinin often produces headache. Anesthetics, aconite, belladonna, carbon monoxid from excessive smoking, caffein, copaiba, digitalis, ergot, lead, mercury, opium, veratrum may produce headaches. Alcohol in excess, or in some people, even in small quantities, often causes headache. This may be produced by the acidity of wines, by the toxic effects on the brain, by digestive disturbances, by the constipating effect, by the fusel oil in cheap whiskey or in chronic alcoholism by chronic thickening of the meninges. Among rarer drugs may be mentioned anilin, carbon bisulphid, sulphuretted hydrogen, strychnin, turpentine. Iron produces headache when given to people who already have enough in their blood; sometimes diuretics, by leading to a concentration of the blood, will produce severe, persistent general headache. The author saw one case, from drinking lithia water, in which the hemoglobin was 120 per cent. but gradually returned to normal after the lithia water was stopped. *Plethora* not uncommonly produces headaches through an increase in the quantity of blood in the body. The author has seen intense headaches in Osler's disease (polycythemia), as well as from polycythemia produced by excessive diuresis, and hyperhidrosis. Suppressed menstruation may also cause plethoric headache.

Blood-vessels.—Arteriosclerosis sometimes causes headache, probably from inanition of the brain. Hypertension may produce headache, but many cases with extremely high blood-pressure never have headaches. The tension usually increases during the paroxysm. The headache is apt to occur during the morning and evening alkaline tide.

Lumbar puncture is often followed by headache if the pressure of the spinal fluid is low, or lowered by the withdrawal of too much fluid, but it may be relieved by elevating the foot of the bed.

Rheumatism of the muscles of the head (*frontalis*) and neck causes severe pain or a dull ache, which is made worse by the movement of these muscles, or by pressure on them. Subcutaneous fibroid nodules may exist on the scalp or back of the neck and be very tender on pressure.

The Sexual Organs.—The influence of the sexual organs in the production of headaches is probable, occurring as follows:

(a) From insufficient internal secretions brought about by nocturnal emissions, spermatorrhea, and allied losses.

(b) From nervous strain, produced by erotic impulses, unsatisfied sexual desire, restlessness at night, dreams, disturbed sleep, etc.—both in the male and female. Block has reported headaches in women due to nocturnal orgasms, and since this paper was written many such cases have come to his attention.

(c) Diseases and displacements, such as uterine fibroids, flexions, versions, erosions of the cervix, suppressed or scanty menses, after labor or abortions, endometritis, etc.

Menstruation.—Monthly recurring headaches may occur in girls before puberty. After the menses begins, the headaches usually occur just before the flow, or in some cases a day or two before; they either cease when the flow starts, or they may last the first day of the menses, but rarely during the whole menses. Sometimes they begin during the flow, rarely after it. Various theories of the mode of action have been advanced. Mackenzie thinks it due to congestion of the nasal mucous membrane, while Haig attributes it to uricacidemia. Head thinks it due to lowered specific resistance of nerve-centers to sensory impulses. It seems probable, however, that swelling of the hypophysis during menstruation may be a cause of menstrual headaches. The whole nervous system is upset in many women at the menses. When a predisposition to headache exists, this condition would easily cause it purely as a nervous manifestation. By some this headache has been supposed to be a reflex headache; by others as due to toxic products from the ovaries.

Monthly epochs occur according to Campbell during *pregnancy* and *lactation*, and headaches may occur at these epochs; or the pregnancy may have a beneficial effect and the headaches cease entirely. During the postclimacteric epochs headache is very common. During the menopause women more frequently suffer from headaches than during any other time; these headaches usually start before the first irregularity, and may last for years after its cessation.

Either amenorrhea or menorrhagia may cause headache; headaches may coexist with dysmenorrhea or irregularities in time. Both of these are nervous in origin, and frequently due to unnatural sexual excitation or anemia.

Kelly found that 107 out of 500 pelvic and abdominal cases suffered from headache; in 32 cases the headache was associated with the menstrual period. "Menstrual headaches are vasomotor in origin." "As a rule, the premenstrual form is relieved when the flow appears, and the menstrual form when a sufficient flow is established."

Headaches are often produced by anemia from uterine hemorrhage. When the headaches follow the menses they are often due to excessive flow.

Pregnancy may relieve or temporarily cure headaches of menstrual origin. But pregnancy may itself cause headaches, which are more common from the third to sixth month, and the tendency to headache increases as the climacteric is approached.

Lactation, especially if prolonged, has a tendency to produce headache from anemia, exhaustion, or prolonged cessation of the menses.

Syphilis.—Syphilis may produce headaches through gross pathological lesions or in other cases through intoxication. In the secondary stage headache is very common, and occurs also in the febrile attacks of syphilis, probably partly from the fever. They are more severe in

women than in men. The organic headaches in syphilis are due to meningitis or gummatous formations or arterial changes, or changes in the skull. The headache often precedes by days or months the paralysis or aphasia from syphilis. It is often localized, and accompanied by tenderness, but in the majority of cases the headache is general, becoming worse in the afternoon and night. When meningitis is present the headache may be continuous and excruciating.

Symptomatology.—**CLINICAL MANIFESTATIONS.**—Of course the chief and essential symptom is an aching sensation in or on the head which may be strictly localized to a small area (clavus), and may remain localized, or may extend from this area and become more or less general. Headache may be unilateral or bilateral. Headaches in the front of the head are spoken of as frontal or sincipital; those in the top of the head as vertical; those in the back of the head as occipital; those in the side of the head as parietal; those back of the ear as postauricular; those in the temples as temporal. The seat of the pain depends upon two factors: (1) a predisposition to pain in a certain part of the head, in which case whatever the provocative causes may be, the pain centers at this point, and may be reproduced at this point by different causes; (2) the determination of the seat of the pain by a definite cause for the headache which is practically always at the same point, occasionally varying (probably from other factors coming into play), or often becoming general when severe, or perhaps being always general and not localized. In general debility, the headaches are usually vertical; in neurasthenia, they are usually frontal, but may be vertical. In hysteria, they are usually general, but may be frontal or vertical. In pelvic disturbances, they are said to be most often vertical. Dyspeptic headaches are usually frontal. In cases which always have pain in the same place, never varying, the headaches are often due to a definite fixed cause, while in those where the pains vary in site and place they are usually functional, and due to a general cause. When the headache is on both sides it is usually more intense on one side than on the other. In other cases, it is entirely unilateral.

Character of the Pain in Headache.—It may be dull and continuous, but rendered worse by coughing, sneezing, exertion, bright light, noise, etc. It may be throbbing, shooting, burning, splitting, etc. The headache may be paroxysmal with freedom from pain between attacks. It usually comes on gradually, or the patient may wake with it. Campbell says, "The painful area (in unilateral headache) may more or less exactly correspond to the distribution of a particular nerve, such as the supra-orbital, supratrochlear, nasal, auriculotemporal, great auricular, or great occipital." A portion of a nerve area or several nerve areas may be involved. The pain usually comes on gradually, though it may be sudden in onset and cessation.

(a) **The Accessory Sinuses of the Nose.**—Pain in sinus inflammation occurs according to Grunwald in 100 per cent. of acute cases, and in 50 per cent. of chronic cases. The pain often recurs at the same time each day, usually in the forenoon, and lasts several hours.

The pain in *frontal sinusitis* is usually limited to the glabella or supra-orbital region, but may affect the whole frontal region. It is rarely occipital and is on the side of the inflammation. There is marked periodicity of the pain, usually from 10 A. M. to 2 or 4 P. M., when it suddenly ceases.

The *ethmoid* usually causes dull pain between the eyes with a feeling of weight in the vertex.

Maxillary sinusitis causes pain in that area but in chronic cases it is often supra-orbital (one-quarter of the cases), and confined to the side of the lesion.

Sphenoidal sinusitis can cause severe pain in the temples, postauricular region, and middle ear, with a feeling of weight and pressure in the vertex. The headache may be the only symptom of the disease. Both the sphenoid and posterior ethmoid cells may cause pain in the occipital region.

The headache from *intranasal disease* (obstruction) is usually at the root of the nose, over the glabella, or extends to the frontal region. The pain is usually worse on waking in the morning; it frequently awakens the patient about four or five o'clock in the morning, when, too, the nose is filled up; it often decreases during the day, and is easy in the afternoon. This form of nasal obstruction is often associated with priapism, which is usually slight and not associated in the mind of the patient with the condition, but may be severe and persistent. Both the headache and the priapism usually terminate on clearing the nose of mucus, on contraction of the swollen tissues by suitable treatment, or by the free discharge of mucus or pus from obstructed accessory sinuses, or epistaxis. The most frequent location of the pain in sinus disease will be seen in the diagram taken from Skillern.

(b) *The Eyes*.—Various disturbances of the eyes give rise to headache, which is most often frontal, sometimes occipital. "According to Culver and Dana, the pain resulting from errors of refraction is generally frontal, while that occurring in consequence of muscular errors is more commonly occipital." (Quoted by Campbell.) Frontal headache is less common among the blind than in those who see well; but the blind are more liable to parietal headache. Campbell suggests the possibility of ear-strain comparable with eye-strain.

When the headache is on one side, it is usually on the side of the greatest ocular defect—such as in anisometropia. There is usually a history of eye-strain, or an occupation which involves excessive or close work with the eyes. The headaches often come on about noon after several hours' work, but on the other hand the patient may waken with the pain in the morning. While the headache is usually frontal, it is sometimes occipital.

Flemming says hypermetropic headache is most often frontal, above each orbit, but it may be occipital and nuchal. "A very characteristic feature is that the patients waked up with headache which improved after breakfast and might be absent during the day if the eyes were not much used, but would be increased or brought about by the use

of the eyes for near work." According to Crawford, eye-strain most frequently causes brow-ache or supra-orbital headache over one or both eyes; next is deep orbital pain (usually astigmatism). Fronto-occipital headache may be most acute in the morning due to eye-strain on the previous day, and occurs especially with astigmatism, with axes deviating from the vertical.

The symptoms of eye-strain, aside from headache, are fatigue on using the eyes, blurred vision, discomfort, or even tenderness of the

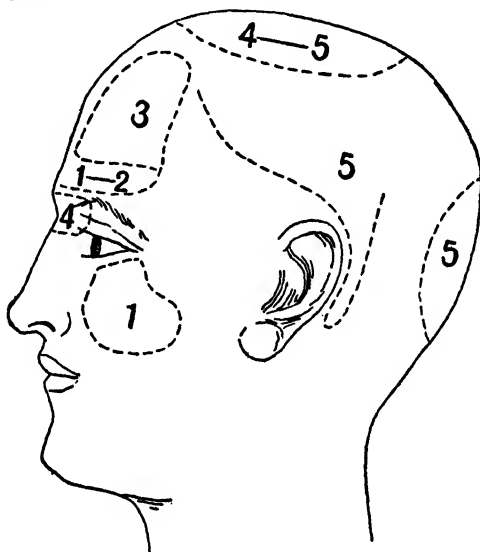


FIG. 1.—DIAGRAM SHOWING THE MORE FREQUENT SITUATIONS OF PAIN IN ACCESSORY SINUS DISEASE. (After Skillern.)

The exceptions, however, are more frequent than the rule. (1) Acute maxillary sinusitis; (2) acute frontal sinusitis; (3) chronic frontal sinusitis; (4) chronic ethmoidal inflammation; (5) chronic sphenoidal sinusitis.

eyeballs; when fixation is defective, mental bewilderment may occur, due to the images received from each eye not being superimposed accurately.

(c) *In febrile headache*, the pain is in proportion to the height of the fever in many cases. On the other hand, certain febrile diseases give rise to more marked headache than others, quite independently of the height of the fever. Thus, in small-pox there is intense frontal headache.

(d) Various *toxic conditions* give rise to general headaches, such as uric acid intoxication, Bright's disease, constipation, and intestinal disorders with their associated symptoms.

(e) The headaches from *organic intracranial diseases* are usually severe and persistent. The headache from meningitis is the most intense known. Headache from brain tumor is at first intermittent, but the attacks tend to be closer together and more persistent, and finally are constant, with acute exacerbations. These attacks probably represent congestion in or around the tumor, or have to do with a variation in the supply of blood to the tumor. Brain abscesses, like brain tumors, may produce severe and persistent headache, which may be worse in one part of the head than in another, but does not always correspond to the position of the tumor, although frequently in cerebellar abscess or tumor, the pain is occipital and nuchal. In brain abscess there may be latent periods in which all pain disappears.

"Whatever tends to aggravate functional headache, such as stooping, a fit of coughing, excess of nitrogenous diet, tends also to aggravate organic headache" (Campbell). Headache, no matter what the cause, is usually made worse by constipation, straining at stool, stooping, sudden jarring, severe mental strain or loss of sleep. When the headache is due to nasal disease it is often associated with impairment of the cerebral functions, defect in memory and concentration.

Other Symptoms Accompanying Headache.—During the headache various other symptoms are present. The patient may be depressed and irritable. The power of concentration and attention is diminished. There is disinclination to work, mental slowness and drowsiness. Photophobia is a common symptom; the patient is usually found in a darkened room, preferring quiet and solitude. Occasionally dimness of vision, deafness and tinnitus accompany the headaches, but usually there is hyperesthesia of sight and hearing. There may be a feeling of numbness and weight or pain in the ears during the headache.

PHYSICAL FINDINGS.—Sometimes there is soreness, pain or tenderness in the eyeballs, or pain on moving the eyeballs. There is a decrease in the luster of the eyes in headache, except in fever. Usually during headache there is enophthalmos and a distressed look, with a pinched, drawn expression and slight blueness of the skin. The hands and feet are often cold to the touch. Sometimes the pain is accompanied by flushing of the face and profuse sweating. If fever be present, the eyes may be bright and prominent and conjunctival congestion may occur.

In some cases the temporal arteries are distended, tortuous, and throb violently (although the face is usually pale during the attacks); this may be only on the side of the headache if it be unilateral.

The flow of urine is decreased during the attack.

Campbell says that in those accustomed to supra-orbital pain he has often noticed asymmetry in the frontal wrinkles and the height of the eyebrow. There may be corrugation between the eyebrows, due to contraction of the corrugatores supercilii, and transverse wrinkling of the forehead due to contraction of the occipitofrontalis, especially if the headache be frontal, orbital or if photophobia be present. There is

partial closure of the eyes due to the sphincters, and drooping of the lids from relaxation of the levatores palpebrarum.

In febrile cases, the face is usually flushed, also in congestive headaches associated with pulsation of the carotids. In non-febrile cases, the face is usually pale and shrunken, and the complexion sallow or bluish. There is a general expression of suffering. In some cases the patients walk the floor in pain, but usually they feel better lying down. The hands may press on or rub the head over the seat of pain, or they may even beat upon the head; and in one of the author's cases the patient thrust her thumb deep into the orbital fossa above the eye till it was feared she would dislocate the eyeball. Sometimes the patient tosses the head from side to side in bed or may cry out in anguish. Children may beat or rub the head, or wear the hair off the back of the head from tossing on the pillow.

In some cases a localized swelling may occur on the scalp or a "Pott's puffy tumor" may appear. Grayness or alopecia sometimes occurs.

In some cases there is a localized elevation of temperature on the scalp which is evident to the sense of touch. Tenderness of the scalp may be present in intracranial disease; in organic disease of the cranium, pericranium or epicranium; in rheumatic or gouty headaches; or from other causes, and may exist without headache being present. Tenderness may only be elicited on examination, or may be very evident to the patient, especially on combing or brushing the hair. It is usually associated with headache, and may precede, coexist, or follow the pain; but the area of pain and tenderness may not exactly correspond. The hairy portion of the scalp, especially the vertex, is most liable to tenderness. In cases of brain tumor the tenderness is more indicative of the seat of the tumor than is the seat of the headache.

Diagnosis.—The diagnosis of the existence of headache rests entirely upon the statement of the patient, except in infants, in whom it may be suspected by their actions (*see* Physical Findings). What is of more importance is the determination of the cause of the headache. *Malingering* is common, and is more common in headache than in all other diseases combined. It is the usual social excuse, and is often used to cover any disinclination on the part of the patient to do other people's ways. But it should not be forgotten that it is often the first symptom of inflammatory or febrile disorders, and the author has seen cases of *appendicitis* which at the time of diagnosis gave no other complaint except headache. The temperature should always be taken in cases of unusual headaches and the various factors given under etiology should be considered.

When due to *nasal obstruction*, inhalations of steam will shrink the nasal mucosa, and increase drainage. If headache is relieved by this procedure, it indicates that the cause lies in the nose, or one of its accessory sinuses. The same effect may be produced by chloretone, camphormenthol, or cocain sprays. By cocainizing different portions of the nose it can be determined which part of the nose is the cause. By irritating different parts of the nose with a probe it can be determined which pain aggravates or reproduces the headache. Inflation of the mid-

dle ear relieves headache when due to the ear. In purulent otitis, when the pain is in the occipital region and side of the neck, it usually indicates meningeal involvement and is often fatal.

Brain tumor and *brain abscess* will usually show some focal evidences, and a gradual march of symptoms and signs, optic neuritis, etc., which should be excluded by examination. Jenner points out that the continuance of pain after vomiting has ceased is strongly suggestive of its meningeal origin and occurs in all forms of *meningitis*. *Cerebral syphilis* may be excluded by its symptoms and signs, or if not, then by the usual blood and spinal fluid tests (Wassermann test, cell-count, and globulin test).

Headache due to the *eyes* can be excluded by the usual eye examinations; but it is suggested by the occupation of the patient, the history of over-use of the eyes, the time of day of the occurrence of the headache, the frontal or occipital location of the pain, etc.

According to Gowers, a headache that prevents sleep is organic, while one which does not interfere with sleep is functional; Campbell thinks this true of minor headaches, but thinks any severe pain may interfere with sleep. Sleep often relieves headache.

Supra-orbital headache always rouses suspicions of *malaria* ("brow-ague") and often occupies a smaller area than the distribution of the supra-orbital nerve.

Occipital and nuchal headache leads to a question of *cerebellar tumor* or *nervous strain*, which are easily differentiated. Sometimes the question of differentiation of *neuralgia* and headache may offer difficulties, especially as tenderness may be present with headaches, and according to Campbell may be absent in neuralgia. But the character of the pain and strict limitation to the area of distribution of the nerve in question are usually sufficient, though it is a question whether headache itself is not often a neuralgic condition of the meningeal branches of the trigeminal nerve.

Leftwich gives the following lists of headaches classified according to the seat of the pain:

Frontal Headache: Adenoids, *anemia*, enteric fever, eye-strain, fevers (prodromal stage), frontal *sinus obstruction*, gastritis, glaucoma, hematoma of dura mater, iritis, lithemia, malaria, neurasthenia, periostitis, syphilitic nodes, thrombosis of the superior longitudinal sinus, trigeminal neuralgia, uremia.

Occipital Headache: Adenoids, *asthenopia*, Buhl's disease, cerebellar tumors, cerebrospinal meningitis, cervico-occipital neuralgia, cirrhosis of kidney, *constipation*, diabetes, epilepsy, eye-strain, gout, locomotor ataxia, nasopharyngeal disease, nephritis (chronic), *neurasthenia*, pharyngitis, rheumatism, sphenoidal disease, *syphilis*, syringomyelia, tongue lesions (basal), uterine diseases.

Unilateral Headache: Adenoids, antral abscess, bone (diseased), cancer of tongue, carious teeth, dysmenorrhea, eye-strain (unilateral), gouty state, *hysteria*, mastoid abscess, *migraine*, nephritis (chronic), otitis

media, **polypus (nasal)**, *trigeminal neuralgia*, tumor (cerebral), *wax in meatus*.

Pain in Vertex: Anemia, chlorosis, *constipation*, epilepsy, *hysteria*, *neurasthenia*, uterine disease.

Unclassified Headaches: Abscess of brain, Addison's disease, ague, alcoholism, *amenorrhea*, *anemia*, apoplexy, arsenic poisoning, asthma, atony of stomach, aura epileptica, catalepsy, chlorosis, cinchonism, cirrhosis of kidney, compression of brain, congestion of liver, *constipation*, *coryza*, dengue, diabetes, dilatation of stomach, disseminated sclerosis, duodenal catarrh, *dysmenorrhea*, dyspepsia, dyspnea, embolism (cerebral), encephalitis, epilepsy, erysipelas, exophthalmic goiter, general paralysis, glandular fever, glaucoma, gouty state, hematoma of dura mater, *hemorrhage*, *hay-fever*, hereditary cerebellar ataxy, hydrocephalus, hyperemia of brain, hypertrophy of brain, hypertrophy of heart, *hysteria*, *incubation of fevers*, *influenza*, iritis, jaundice, lactation (prolonged), lead poisoning, leontiasis ossea, leukorrhea, lithemia, measles, meningitis, menopause, mental over-strain, morphinism, nephritis, *neuralgia*, *neurasthenia*, oöphoritis, oxaluria, pachymeningitis, plague, pneumonia (acute), polycythemia (splenomegalie), polypus (nasal), pyrexia, relapsing fever, remittent fever, *rheumatism*, softening of brain, spur of septum, sunstroke, syphilis, tapeworm, tension (high arterial), thrombosis (cerebral), tumor of brain, turbinated bone (enlarged), typhus fever, uremia, valvular disease, variola (first stage), Weil's disease, impure air, fatigue, flatulence, depression after excitement, and imperfect coagulability of the blood. Persistent severe headaches always suggest organic brain disease, such as tumor, abscess, cerebral syphilis, meningitis, etc.

The family relations, general happiness and conduct, occupation, sexual habits, dreams, mode of living, ventilation, sleep, proper use of the pillow, eating, bowels, hereditary influences, and various other factors which are not shown by physical examination should be considered.

Association with Other Diseases.—Headache is always a symptom, never a disease. So-called periodic sick headaches are considered under migraine. The diseases with which headache is associated will be found under diagnosis and etiology.

Headache from eye-strain may be associated with nervous disorders, such as vertigo, palpitation, vomiting and neuralgia.

Clinical Varieties.—*Cephalalgia hemorrhoidalis* is a variety of headache which occurs in plethoric people who have periodic bleeding from hemorrhoids, or headache when this bleeding fails to occur.

Cephalalgia pharyngotympanica has been described by Legal as due to aural and pharyngeal disease (Legal's disease).

Academy headaches are from strain of the occipital muscles in visiting picture galleries, motion picture shows, museums, etc.

"*Bad husband headaches*" of Frances Cobbe are produced by emotional disturbances, neglect, crying, and marital unhappiness.

Hysterical headaches are due to emotional disturbances, desire for sympathy, malingering, etc.

Cephalalgia puerperalis, due to postpartum hemorrhage, may occur first a few days after the hemorrhage, is severe in nature, and lasts for several days or weeks.

Cephalalgia e vino is caused by excessive drinking, mixed drinks, and especially whiskey, when fusel oil is not distilled off from it.

Cephalalgia degenerativa occurs in neurotics, or in people with hysteria, neurasthenia, etc.

Cephalalgia catamenialis is a form of headache occurring at the menstrual period.

Cephalalgia gravidarum is apt to occur about the third month of gestation.

Clavus, clavus hystericus, galæ, monophagia, is a form of headache in which there is a piercing pain in a limited area. The pain may be supra-orbital, occipital, parietal, or in the vertex, and may cause nausea and vomiting; it may occur at the same hour each day, and much resembles migraine and neuralgia. It occurs especially in anemic and debilitated patients. The sensation is that of a nail being driven into the head.

Cephalæa adolescentium (of Charcot) is probably of sexual origin. Among the older writers *soda* meant headache in general, while the term *cephalæa* was used to indicate pain in the whole head; *hemisrania*, pain in one side of the head; *clavus* or *monopagia*, pain in a small area; and *ovum* or *dolor fortis galeatus*, used to denote pain in the top of the head, like a helmet.

Treatment.—TREATMENT DURING THE ATTACK.—The patient should be kept quiet in bed in the majority of cases if the headache is severe. Some cases find more comfort in walking constantly. In milder cases rest is not necessary. In severe cases where there is hyperesthesia the room should be dark and quiet.

Many cases are relieved by analgesics, such as **acetyl salicylic acid** (grains v to x [0.324 to 0.65 gram] every 4 hours), **acetanilid** (grains ii to iv [0.13 to 0.26 gram] every 4 hours), **pyramidon** (grains v [0.324 gram]), **phenacetin** (grains v to x [0.324 to 0.65 gram] every 4 hours), **anesthesin** (grains iii [0.195 gram]), **trigemin** (grains vii ss to xv [0.49 to 0.97 gram]) and other analgesics mentioned in the treatment of migraine. Great caution should be used in giving such drugs as acetanilid, etc., during menstruation, as they are particularly toxic at this time and often give rise to cyanosis, dyspnea, and cardiac weakness. During a headache the stomach often ceases to absorb anything; if several doses are given during a headache, they may all be absorbed at the same time when the headache terminates, thereby leading to toxic results. It is usually better to protect against the depressing influence of these drugs by the combination with **caffeïn** (alkaloid grain ss [0.324 gram] or citrate grains iii [0.195 gram]) or with **strychnin sulphate** (grains 1/40 to grains 1/30 [0.0016 to 0.0021 gram]).

When the headache is due to the eyes, or nose, lying down with a **hot-water bag** over these regions for half an hour or an hour will often give relief to the patient. In nasal obstruction, **inhaling steam** or a

cocain spray gives immediate relief; a drop of **cocain** in the eye will relieve a headache due to eye inflammation. When the headache is due to the ear, **cocain** in the ear, a **hot-water irrigation**, or **hot-water bag** is often helpful. **Hot or cold applications** to the head often give relief. Rubbing with **spirits of camphor**, a **menthol cone**, or **chloroform**, often gives much comfort. **Mustard plasters** may be applied to the back of the neck or temples, and sometimes a **fly blister** on the temple will give prolonged relief when the pain is located there. Gentle, or in other cases deep, rubbing or **massage** of the head or scalp does good.

In severer cases **morphin** (grains $1/8$ [0.0081 gram]) hypodermically may be necessary, while in hysterical cases the **injection of sterile water** usually suffices. **Infiltration anesthesia** by Schleich's method is valuable during the attack, or injection of the painful points with 0.2 per cent. salt solution. In plethoric cases **bleeding** may give prompt relief.

Roger and Baumel found that **lumbar puncture** gave relief in headaches due to acute infectious diseases, in a few minutes after puncture. In almost all (of 15 cases treated) there were hypertension and excess of albumin. In cases of meningitis I have found the extreme headache relieved by lumbar puncture before the needle was withdrawn. Gordon found that lumbar puncture gave excellent results in relieving headaches in typhoid fever, pneumonia, and influenza. He removed 15 to 40 c.c., depending upon the tension. He found the fluid sterile in all of them and the tension high. Some showed leukocytosis and others an increase of albumin in the cerebrospinal fluid. When the pain is located in the back of the head and neck and is due to fatigue or nerve-strain **cafein** is very valuable.

Brunton finds that frontal headache with constipation is relieved by **magnesium sulphate**; headache just above the eyebrows without constipation, by **acids**; headache higher up without constipation, by **alkalis**. In cases with hyperacidity of the urine, alkalies should be used. In cases due to Bright's disease or diabetes the treatment should be directed to these diseases. Arteriosclerotic cases are treated by **diet** and the **iodids**.

TREATMENT BETWEEN ATTACKS: TREATMENT OF THE CAUSE.—The treatment, as far as a cure of headache is concerned, depends entirely upon its *cause* for permanent relief. The various etiological factors should be considered and corrected.

In some cases no cause can be found and the treatment must be directed to general **hygienic measures**, and the relief of pain. Even when the cause is found, it is necessary to find *the cause of the cause*. Thus, in the treatment of asthenopia or heterophoria, it should always be borne in mind that the mere wearing of glasses is often not the essential point at all, but that these conditions are often directly due to some condition of general health which, when corrected, accomplishes the cure of the eye trouble and consequently of the headache. In one of the author's cases with adductor weakness this was due to anemia, which in turn was due to a mild tuberculosis; the headache and eye trouble disappeared after improvement in the general health.

In *eye cases* Walton thinks no case is completely tried until (a) the glasses are properly centered; (b) their continued readjustment is practiced; (c) the patient looks as much as possible through these centers instead of from side to side; (d) efforts to strain the eyes to see distant objects with the glasses are avoided; (e) spectacles instead of eye-glasses are used; and (f) the use of spectacles is constant, not intermittent. **Atropin**, by paralyzing accommodation, will relieve headache in appropriate cases.

In *anemic cases* iron and arsenic are valuable.

In *toxic cases* laxatives and diuretics should be used. **Diuretin** has been advised in arteriosclerotic cases (grains v to xv [0.324 to 0.97 grams] t.i.d.) and probably acts by removing fluid from the blood-vessels, thus lowering the tension. When the blood-pressure is high the fluid intake should be restricted.

Uterine diseases and displacements should have appropriate attention.

Diet.—Diet is necessary in cases of *chronic constipation* or when *intestinal intoxication* exists; also in *gouty*, *rheumatic*, *diabetic* and *nephritic cases*.

Climate.—A change of climate is often valuable, especially in cases where the patient is *over-worked and worried*, and, therefore, needs relaxation. It does not seem to matter much where the patient goes, but it is advisable to try a climate opposite to the one in which he lives. Cases which sleep poorly will do better at the seashore.

Hydrotherapy is valuable in *toxic*, *rheumatic* and *gouty cases*. **Hot foot-baths** or **mustard foot-baths** are helpful in congestive headaches, and in menstrual headaches when the flow is insufficient; they should never be used when the flow is excessive. **Cabinet baths**, **sweat baths**, **hot packs**, **hot baths**, and **large quantities of water** are indicated.

Organotherapy.—In *neurasthenic cases*, and especially those of *sexual origin*, **testicular substance** is especially valuable. When the menses are excessive, **mammary substance** should be used in doses of two tablets thrice daily between menses and six tablets thrice daily during the menses. According to Block, it reduces both the quantity and frequency of menstruation. When the menses are deficient, **ovarian substance** and **hot foot-baths** are helpful.

Psychotherapy is very helpful in *hysterical cases* and *malingering*.

Electrotherapy is helpful in *hysterical* and *neurasthenic subjects*, probably from suggestion.

In *syphilitic cases*, **iodids**, **mercury** and **arsenic** are indicated. The iodids soften gummata and give the mercury and arsenic an opportunity to kill the bacteria.

Ext. Ergot (Ext. grains ii [0.13 gram] t.i.d. or F. E. m xx [1.3 c.c.] t.i.d.) is especially helpful in cases of *hysteria*, and in headaches due to *spermatorrhoea* and *nocturnal emissions*.

Setons are rarely used in late years, but they cured the only case in which the author has tried them.

Surgical.—Particularly in nasal obstruction and accessory sinus disease are operations for the relief of headache indicated. Thus the sub-

mucous resection of a deviated nasal septum, or removal of the middle turbinate bone, etc., may relieve long-standing cases of headache when due to this cause. The drainage of an inflamed sinus, the removal of adenoids or diseased tonsils, attention to the teeth, are all of great importance. Cicatrices and depressed fractures may require attention, or in acromegalic headaches, the removal of the pituitary body.

General Management.—Some cases are greatly benefited by daily systematic **exercise**, and by **sleeping on a porch**. In all cases the **bowels** should be kept reasonably well open, and in toxic cases an occasional dose of **calomel** should be given.

Prognosis.—The prognosis of headache depends entirely upon the cause and the ability to relieve it.

Headaches are rare in old people. If they existed previously they tend to become milder or to disappear. Headaches developing in old age are most often due to nephritis or arteriosclerosis, while those beginning at about 40 years of age are most frequently due to presbyopia. Nasal disease often causes impairment of the cerebral functions, defects in memory, and concentration. Gout is often preceded by headaches for many years, but after the gout is fully developed the headaches are uncommon.

Violent and persistent headache may lead to hypochondria.

The frequency of the headache depends upon the cause, and the duration is very variable. In some cases, as in the onset of a febrile disease, it may be an isolated occurrence in the life of the patient; in others there may be a daily headache, as from eye-strain or nasal obstruction, or a steady, persistent dull ache with severer attacks from time to time, as in brain tumor. The prognosis is, of course, that of the etiological factor. Idiopathic headache is often permanent, though not continuous.

Pathology.—It has not been definitely proven what exact structures are involved in headache, but it is probable that many different structures may be involved. The brain itself is said to be insensitive to pain. The meninges are, however, highly sensitive to pain; and this may be provoked by pressure upon the meninges, as by a tumor, or by congestion from vascular disturbances, acute or chronic inflammation or thickening, with pressure upon the nerves which pierce it. Disease of the bony portion of the skull, the periosteum, the pericranium or epicranium are all at times the seat of headaches. Small apertures exist in the aponeurosis of the cranial muscles, in the periosteum of the skull and in the skull itself, through which blood-vessels and nerves inside and outside of the skull are closely related, and very little swelling suffices to produce pressure upon them. The pericranium is continuous with the dura mater through these apertures, the sutures, and through the orbit and sphenoidal fissure, and the latter is continuous with the sclerotic of the eye. The holes in the aponeurosis of the occipitofrontalis are, according to Campbell, less yielding on top of the head; there is, therefore, greater liability to pressure on nerves and blood-

vessels here, than nearer its margins where the apertures are loosely woven.

The front half of the head is supplied by the fifth nerve, the posterior half by the occipitals, great auricular and Arnold's nerves.

Thickening of the cranial bones, as well as the meninges, has been found in chronic alcoholism, chronic renal disease, and general paralysis of the insane. Direct involvement or pressure upon the bones or meninges are probably responsible for the pain in tumors, abscess, syphilis and hydrocephalus. The intermittent character of the headache in brain tumor is probably accounted for by variation in its blood supply.

Febrile headaches probably produce pain through congestion. The mode of production of toxic headaches is uncertain.

Sociological Aspect.—The relation of headaches to marriage depends upon a careful consideration of the individual case and the etiology of the headaches. In some cases the headaches are so severe and frequent that they interfere with plans of life and no engagements can be definitely made. In some cases the influence on the offspring must be considered, as headaches and the etiological factors of headaches may be inherited from one generation to another. As a rule, however, headaches, unless extreme, should not interfere with marriage.

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CHAPTER XIX

MIGRAINE

(*Sick Headache or Periodic Headaches*)

By E. BATES BLOCK, M.D.

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Synonyms.—The term *hemicrania* should not be used for migraine as it is also applied to cranioschisis and is thus confusing. *Megrim* is often used in England, though rarely in this country. *Emigranea* and *semicranea* are also occasionally used. *Periodic sick headaches* or *bilious headaches* are the terms most used by the laity.

Definition.—By migraine is meant periodic attacks of pain in one or the other side of the head, often associated with vomiting and followed by sleep.

While the above definition is usually true, it may be stated that headache may be on one side of the head only, or upon both sides, while migraine, though usually on one side of the head, may be on both (Gowers, Campbell, and others). It is, therefore, apparent that there is no clear differentiation between headache and migraine, in so far as the location is concerned; and we may also say that the etiological factors are for the most part the same in the two "conditions" with some exceptions. All cases of migraine are cases of headache, but not all cases of headache are migraine. There is no difference except in the extent of the seat of the pain between *migraine* and *periodic sick headaches* and they will be treated as the same disease.

Etiology.—**PREDISPOSING CAUSES.**—*Time of Life.*—The disease often begins in early childhood; patients frequently use the expression "as long ago as I can remember." The vast majority begin before twenty-five years of age, although it may begin as late as fifty or sixty years. It often begins at puberty, especially in girls. Alger finds 3 per cent. of cases develop between the ages of five and ten years.

Time of Day.—The attacks may start any time of the day, but they usually begin early in the morning on waking, or soon after waking. Rohrer found them four times as often in the daytime as at night in his cases.

Occupation.—The disease is rare among the uneducated poor. It occurs in people of neurotic family history, especially in those who lead a studious life.

There is no apparent influence of seasons of the year.

Sexual Disturbances.—*Menstruation* has a marked influence on the attacks. They occur most frequently just before the menses, less often during it, rarely immediately after it. The *menopause* usually has a favorable influence but occasionally provokes attacks. Menstruation and migraine often start at the same time, or the onset of menstruation may provoke a previously existing migraine. Migraine has been known to follow *hysterectomy*.

Migraine is said to occur as a reflex from diseases of the *sexual organs*. Levi thinks the attacks of migraine which occasionally accompany the menstrual periods may be due to auto-intoxication of ovarian origin.

Pregnancy may cause a cessation of migraine or it may come on after pregnancy.

Among the exciting causes of the attacks Oppenheim mentions mental exhaustion, prolonged emotion, work in overheated rooms, possibly onanism, sometimes coitus, excitement, alcoholic excess, bad air; and Wallis mentions errors in diet, worry, overwork, fatigue, late hours and uncorrected ocular defects.

Sex.—Females are decidedly more often affected than males. In the 23 cases reported by Sapogenix, 5 were men, 15 women and 3 infants.

Heredity.—The chief cause of migraine is hereditary predisposition—most often there is similar heredity, sometimes dissimilar heredity. It occurs especially in nervous people or members of neurotic families. According to Möbius it is directly inherited in 90 per cent. of cases. Oppenheim mentions its transmission through four generations, eight members of one family being affected. In one of the author's cases the mother and nine of her mother's siblings had migraine (11 cases in the family). Clark (quoted by Osler) reported recurring motor paralysis in 11 members in 3 generations in one family. It is usually inherited through the mother, and affects the girls in a family more often than the boys.

According to Fuchs, the great majority of cases of astigmatism are congenital, and it is apt to be transmitted by heredity. Hypermetropia is a congenital condition which, however, tends to better itself in the growth of the child; myopia is rarely congenital, but there is an hereditary predisposition. Although these conditions often coexist with migraine and may be causes of it, they are also evidences of hereditary defect.

Syphilis.—The rôle of syphilis in migraine has not been fully studied. In all cases that occur late in life without other apparent cause it should be suspected, as well as in congenital hereditary migraine. According to Halban, migraine may be the only manifestation of hereditary syphilis. In 100 cases of syphilitic disorders of the nervous sys-

tem (without selection), Collins found 2 cases of migraine, one of them associated with ophthalmoplegia.

Nasal polypi, nasal calculi, stenosis due to hypertrophy or swelling of the turbinates, etc., may cause migraine. Diseases of the nasopharynx are recorded as causes.

Organic brain diseases, such as brain tumor, often produce symptomatic migraine.

Thyroid instability, too much or too little thyroid secretion, may, according to Sapogenix, cause migraine.

Diet.—Certain foods may provoke attacks in people who are subject to migraine. In gouty or rheumatic subjects, the proteids are usually held responsible. According to Gowers, in cases of migraine due to errors in diet the error will not produce an attack immediately after one has passed off, but will do so after an interval has elapsed. Digestive disturbances are said to produce migraine. Both achylia and hyperchlorhydria certainly are associated with headaches.

Auto-intoxication is one of the oldest and best known theories and chronic constipation has been frequently observed in migraine. Hunt thinks the cause of migraine attacks lies in the periodical occurrence of an auto-intoxication affecting the sympathetic and vascular systems with localized vasomotor spasm, or vasomotor dilatation of the cerebral circulation (angiospastic and angioparalytic types of migraine). Marked pallor of the optic nerve and retina has been frequently noted in the attacks, as well as variations in the size of the temporal artery. These changes probably occur also in other parts of the brain and account for the scotomata, paresthesiæ, paresis of the extremities, and aphasia. Over-indulgence in nitrogenous food is a frequent cause of migraine. Haig places special stress upon uric acidemia as a cause. He has shown that for several days before an attack of migraine, the uric acid excretion is reduced below normal, while during the attack it rises above normal, due to the elimination of uric acid previously stored up. During the headache the pulse is usually slow, the tension high, the excretion of urine is decreased owing to arterial constriction, and there is a feeling of chilliness with cold hands and feet.

Decomposition of the contents of the stomach is one of the causes given for auto-intoxication. Chronic nephritis is given as a cause of migraine. There is no question that it often produces headache. *Alcohol* or *tobacco* may provoke attacks in those predisposed to it.

Gout.—Gowers says that in the worst cases of migraine, an ancestral history of gout is rarely absent.

Walton regards migraine as an *occupation neurosis* which involves (1) the visual centers; (2) the centers of accommodation; (3) the intrinsic and extrinsic muscles of the globe; (4) the muscles outside the orbit which are called into play in the effort for accurate vision, principally the corrugator supercilii and the occipitofrontalis, and also the muscles inserted in the occipital region, which serve to steady the head.

Alexander says migraine is a vascular disorder affecting various parts of the cortex and the dural branches of the trigeminus nerve.

According to Fuchs some of the cases of ophthalmoplegic migraine are hysterical.

Rohrer compares migraine to *anaphylactic shock*, in which there are sweating, nausea, vomiting, excited intestinal activity, and occasional hemorrhages. Anaphylaxis is inherited only through the mother, and there is a great preponderance of mother heredity in migraine.

Planec supports Charcot's opinion that there is a primary disease, ophthalmoplegic migraine. Möbius considers it always secondary to some organic intracranial disease. Fisher speaks of transient ophthalmoplegia externa associated with attacks of severe *headache*. He does not consider these cases of true migraine, and finds the pain in any part of the head.

Eye-strain.—The literature is full of the influence of eye-strain on migraine. Thus Wallis reports cases of hypermetropic astigmatism, mixed astigmatism, and presbyopia, myopic astigmatism and hyperphoria, with relief from proper glasses. According to Campbell astigmatism is the commonest refractive error in producing migraine. When a patient is blind in one eye and has a defect in the other, the migraine is over the latter.

According to Alger, migraine rarely ever occurs in old age when accommodative power has practically disappeared. The three commonest ocular troubles causing migraine are: (1) Over-use of the ciliary muscle in accommodation, such as occurs in hyperopia and astigmatism; (2) conditions in which binocular vision is impossible without undue strain of the extrinsic ocular muscles; (3) cerebral fatigue that comes from the continual interpretation of distorted retinal images, such as are present in astigmatism and anisometropia.

It is doubtful if presbyopia is ever a cause of migraine, although a frequent cause of headache.

According to Levi, migraine has been supposed to be due to a spasmodic (Dubois-Reymond) or a paralytic (Möllendorf) affection of the sympathetic system, or to be a neuralgia of the trigeminal nerve (Brissaud). Levi thinks the discharge is from a center of the trigeminal fibers in the medulla, and spreads to neighboring nuclei, giving rise to the various other symptoms. The excitation of this center may be emotional, visceral, or due to auto-intoxication.

Charles thinks that the clinical features of migraine may be explained satisfactorily as due to periodic enlargement of the *pituitary gland*. He explains the headache, nausea and vomiting as the result of increased intracranial pressure; the disturbance of vision (mainly hemianopsia) from pressure, and the fortification figures, etc., from irritation of the optic tracts; the vasomotor phenomena from increased secretion from the posterior lobe or from pressure on sympathetic fibers; and the sensory and motor symptoms from pressure on the crus. Plavec takes the same view, and thinks that in migraine it is probably due to active hyperemia and in ophthalmoplegic migraine to a venous stasis which undergoes periodic exacerbations. He suggests a partial lateral

dislocation of the hypophysis in the latter cases, thus accounting for the pressure on the oculomotor nerve in the cavernous sinus.

Cushing found that during pregnancy the pituitary body shows hypertrophic changes with cellular hyperplasia and in multipara fleeting bitemporal hemianopsia may occur in the last weeks of pregnancy. If we look upon menstruation as a "little parturition," this may help to clarify the etiology of headache, but hardly of migraine, since no cases of bitemporal hemianopsia have ever been observed with migraine, and the theory does not cover such cortical manifestations as aphasia, astereognosis, etc. Amenorrhea is an early symptom of hypophyseal disorders, whether in over-function or under-function, but the absence of polyuria, temperature disturbances, changes in carbohydrate tolerance and growth disturbances in migraine are not in favor of a relation existing. It is, however, not improbable that the headaches during menstruation are due to periodic temporary enlargement of the pituitary body.

Symptomatology.—**FREQUENCY.**—The attacks may occur two or three times in a week; or every one, two or three weeks; most often once a month; and sometimes less frequently. The attacks may show a remarkable periodicity so that a patient may be able to predict fairly well when the next one will occur; in other cases there is no regularity about the occurrence of the attacks. Periodicity is especially noticed in attacks which occur with each menstruation. During pregnancy migraine of menstrual origin may cease entirely.

As to the frequency of hemianopsia in ophthalmic migraine, Sopa-genix found it in eight out of twenty-three cases.

CLINICAL HISTORY.—*Prodromata.*—The day before the attack the patient usually has an abnormal appetite, a morbid hunger. Whether or not he gratifies this appetite, the headache follows the next day, but the attack is often less intense if he diets carefully. Often the attack is preceded by constipation, mental depression, or he may feel languid and heavy, with unusual drowsiness the night before the attack. On the other hand, patients often feel unusually well the day before the attacks.

(a) *Sight.*—The prodromata of sight may consist of dark spots, or bright lights which may assume varying shapes such as zigzags or fortification figures, changing in color, shape, position, or size as they develop. They do not occur in the center of vision, but to one side of the fixing point, and are on the side opposite to the headache; or, in cases where the headache is bilateral or medial, they are on both sides. These may always be the same in a patient or may vary in different attacks. The lights may be white or colored, yellow, blue, or red. The visual hallucinations are spoken of as teichopsia. In some cases the lights occupy a segment of the field of vision (scintillating scotoma) in which bright spots, balls of light or colors are seen (irritation), or the vision may be blurred or dim, or dark spots may occur (inhibition). Sometimes there is a shimmering appearance similar to that from the glare of sunlight on water.

In Alexander's case the prodromata consisted of a homonymous
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quadrant hemianopsia to the *right* and occasionally white scintillating scotoma, with pain on the *right* side of the head.

According to Rohrer, the scintillating scotoma is on the two homolateral halves of the field of vision and are equally frequent on the right and left side.

(b) *Cutaneous Sensations*.—These never consist of pain or temperature disturbance. The most common seat is the hand or arm: it starts with a tingling sensation in the fingers, or may gradually pass up the arm, leaving behind numbness and a lessened sensibility to touch, and some weakness; rarely it affects the trunk, and then the leg, but it is often felt in the face, gums and tongue, on one side, but may involve the lips on both sides (Gowers). Its march is slow, never less than five minutes, lasting ten or fifteen minutes. If both the arm and visual symptoms coëxist the former usually precedes the latter, but not always. Very rarely there are cramps or spasms of the muscles on the (peripheral) affected side (Osler). The numbness may affect both hands; it never affects the foot, and rarely the lower limb. It never passes downward, but always upward.

(c) *Cortical Phenomena*.—The prodromata are referable to the cerebral cortex and are in the nature of hallucinations; the side of the cortex involved is the side of the headache. When the headache is on the left side it is often preceded by, or associated with, aphasia. When the cuneus is involved there may be visual hallucinations or homonymous hemianopsia; but very rarely there is a transverse hemianopsia with a loss of vision in the upper or lower half of the combined fields (Gowers). In other cases a scotoma may occur. Hemiparesis and hemianesthesia may occur, and may be permanent. Sometimes the aura consists of vertigo, with or without other disturbances.

(d) *Mental and Psychical Disturbances*.—Rarely mental attacks occur, psychical disturbances, excitement, confusion or depression, which may persist until the attack ends. According to Guidi the mental symptoms may precede the pain one or more days and may consist of manic or depressive symptoms. He found these premonitory symptoms marked in 6 out of 38 cases of migraine and less pronounced in 18 cases.

Mode of Onset.—The attack may come on at any time, but usually starts on waking in the morning. Some cases have no prodromata immediately preceding the attack, the patient simply awaking with pain, while a minority have certain definite symptoms preceding the pain, which is usually the same in each attack with each patient, though it may vary with different patients. These prodromata are sensory in nature and usually involve sight or touch, neither smell, taste nor hearing being affected.

Immediately before the attack there is often a feeling of fatigue, exhaustion, drowsiness, a tendency to yawning, fullness in the head, giddiness and depression.

With or without prodromata, the patient begins to suffer *pain* in one or both sides of the head, varying in location in different patients but usually in the same place in each patient. The pain may be preceded

by slight tenderness which gradually increases as the pain increases; usually it starts with pain associated with tenderness. The pain may start in a small circumscribed area (clavus) and gradually spread; or it may start in its total area at the same time, and gradually increase in severity until it reaches its maximum; when severe, it is attended by nausea, and, when severest, by vomiting, which may mark the crisis of the headache, which then gradually subsides, leaving a feeling of soreness behind, with tenderness. In other cases retching may last for hours after the vomiting and be very distressing to the patient, and the vomiting may not afford any relief. Finally the patient goes to sleep and awakes free from pain. The duration may be only a few hours, but is usually one day, rarely as long as four days.

Subjective Symptoms.—*Nausea* usually precedes the vomiting and does not generally begin until the headache has lasted some time (one to five hours). Retching may be persistent before vomiting occurs, or may last after the vomiting and produce great exhaustion. The vomiting always gives some relief, and may terminate the attack. There is usually a gastrosuccorhea with the attack, with cessation of absorption, so that the stomach may be very full, and at the same time enterosuccorhea may occur with profuse diarrhea. According to Campbell, during the attack there is usually a decrease in the urine, saliva, sweat, and gastric juice, while the attack is often followed by the passage of a large quantity of urine, and there may also be an increase in saliva and sweat. During the attack secretion of tears and sweating are unusual symptoms.

During the attack there is loss of appetite and flatulence. There is cutaneous hyperesthesia of the scalp. Light and noise disturb the patient; the room is darkened and the patient often prefers to be alone. Pain is intensified by moving the head or eyes.

Paresthesia may occur in one arm and is said by Oppenheim to occur sometimes in one or both sides of the body, but anesthesia is rare. In some cases there is anesthesia in the area of distribution of the fifth nerve (Knapp). A weakness in one arm or one side of the body, or even paresis may occur on the side of the body opposite to the headache. In some cases hemiparesis may alternate with migraine. Vertigo, which is often associated with migraine, may exist in some people without the pain.

The crisis usually ends in sleep. Soreness may persist for several days after the attack or transient pain may occur from coughing or sneezing.

Slight attacks may be arrested by occupation, eating, etc.

The attacks which each patient has are not always identical and show great variability in the symptoms in different attacks or may show the same sequence of symptoms in each attack.

Oppenheim says true migraine may be replaced by violent pain in some circumscribed part of the trunk or in an extremity, which after lasting for some hours or even for a whole day, disappears spontaneously, returning in the same way a few weeks later. Albutt finds that gas-

tralgia and migraine do not often coincide in time, but rather alternate the one with the other.

Mania, melancholia, or acute confusion may replace attacks of migraine; or anxiety, dread, fear or depression, may occur with migraine. It may be accompanied by mental confusion, loss of memory, or in other cases the mind is unusually clear and active (Osler).

Forli, in a study of 185 cases of migraine, found the psychical disturbances, though usually slight, are not rare; they may occur one or two days before the attack, or may immediately precede it, constituting an aura, or may occur as the pain is subsiding. Usually they begin at the acme of the attack and gradually subside. Hallucinations of sight and hearing are common, but they exercise no influence on the conduct of the patient.

Gordon reported 12 cases of psychoses developing at the climax of the headache, which disappeared with the headache. The symptoms were confusion, mild stupor, hallucinations (mostly visual), unsystematized delusions, and delirium.

In one of the author's cases there was sometimes a confused delirium during the left migraine attacks, in other attacks motor aphasia, but usually without either of these symptoms. Agraphia or word deafness rarely occur and if so, occur only in left-sided migraine. Aphasia and hemianopsia may be associated. These symptoms usually precede the pain.

Atypical attacks of migraine may occur in which the headache is replaced by nausea (ninth nerve) or vomiting (tenth nerve) or vertigo (Deiter's nucleus), and in the same cases these symptoms may coexist in other attacks with actual pain. Especially in cases of eye-strain may attacks of vomiting occur without or with the headache and particularly in myopic astigmatism and hyperphoria are we apt to have attacks of giddiness and nausea without headache.

Rohrer's description of his case was as follows: The attacks came mostly in the morning. They began with a scintillating scotoma, bow-shaped, which traveled outward in the field of vision, and in half an hour became an ordinary scotoma which lasted five or ten minutes. The sensitiveness began during the scintillating scotoma with a light burning feeling in the conjunctiva. Soon after disappearance of the scotoma, contralateral unilateral headache began. About an hour later, nausea, sweating, and vomiting occurred. He had 250 to 300 attacks in eighteen years.

Campbell says that in 22 per cent. of cases of migraine the pain is worse when the patient is lying down. This occurs especially in cases in which the body and limbs are chilly, and the radials contracted, while there is strong pulsation of the carotids and temporals.

Location of the Pain.—In ophthalmoplegic migraine the pain is usually in the temple. In other forms of migraine the pain may be frontal, parietal, postauricular or occipital; but the relation between the location of the pain and the cause has not yet been definitely established. In the cases of headache due to focal lesions, such as sinusitis, the pain

is on the same side as the lesion, but such causes are more frequently found in unilateral *headache* than in true *migraine*. The same may be said of eye diseases, i.e., the headache when unilateral is on the side of the eye defect, or in cases of anisometropia the headache is on the side of the greater defect. According to Campbell in *migraine*, the right side is affected 7 times to the left 11 times. In mastoid headache, the right side is affected 2 times to the left 5 times. In parietal clavus, the right side is affected half as often as the left. In the occipital region, both sides are usually affected, though one may be worse than the other. Murphy finds that in neuralgic headache involving the suboccipital nerve in women, it occurs on the left side in 99 per cent. of cases, and in functional earache, two-thirds are in the left ear. The left side of the head has been affected much more frequently in my cases than the right. As to the seat of the pain, Campbell says it is wholly extracerebral, and chiefly extracranial. The brain substance itself does not seem to feel pain, but the meninges, the pericranium, and epicranium are all very sensitive, as well as the scalp itself; these seem to be the chief seats of the pain. At whatever point a patient has *migraine*, it is usually in the same place in every attack. Thus one of the author's patients has *migraine* on the left side in 90 per cent. of his attacks, on the right side in 10 per cent. of attacks, while in a very few attacks, it has been upon both sides. In another case the headache is situated in the left posterior parietal region, no matter whether it is brought on by bright lights, loud noises, excitement, over-exertion, eye-strain, emotion (crying), or, in one instance, by postpartum hemorrhage; the pain was in each instance in the same place.

Objective Symptoms.—There are usually no objective signs between attacks except in such types as ophthalmoplegic *migraine*, and often no signs during the attacks, other than pallor, the anxious expression of the patient, and the evidence of pain. In some cases there is permanent narrowing of the palpebral fissure and pupil on the side of the *migraine*; or spasm of the orbicularis oculi may occur during the attack. Oppenheim found sensitiveness to pressure of the superior sympathetic ganglion in many cases. In one of my cases, there was persistent elevation of the left eyebrow with occasional muscular twitching or slow, lazy contractions and relaxations between the attacks of left *migraine*.

PHYSICAL FINDINGS.—Gowers did not observe any changes in the *retinal vessels* during headache. Mollendorf found that "during the attacks the background of the eye on the suffering side was bright scarlet red, the optic papilla red and edematous, the arteria and vena centralis retinæ enlarged, the latter knotty and very tortuous." According to Osler, the retinal arteries are sometimes seen contracted.

Protracted pain may cause dilatation of *blood-vessels in the painful area* and eventually exudation, organization and thickening, and even ecchymoses have been reported.

In temporal *migraine*, the temporal artery on the affected side is often tortuous or may be contracted and small. A slow pulse may occur at the height of an attack of *migraine*.

Grayness or localized *alopecia* may occur on the side of the pain.

According to Oppenheim *albuminuria* and *pain in the region of the kidneys* has been recorded, and also conjunctival and retinal hemorrhage and nasal hemorrhage have been observed.

Sympathetic disturbances may occur, such as pallor at the beginning of the attack, with coldness and sweating of the hands and feet, contraction of the temporal artery, dilatation of the pupils and increased flow of saliva; in other cases the face and conjunctivæ become red, the arteries dilated, and the pupils contracted (Oppenheim); and both of these groups of phenomena may occur in the same attack. Pallor is usual at the onset of the attack. Unilateral hyperhidrosis may occur from sympathetic irritation. The pupils in migraine may be either dilated or contracted, depending on the state of the sympathetics. Sometimes the pupil on the affected side shows alternating dilatation and contraction (hippus). The eye on the affected side is often sunken, and the upper lid droops, and often there is corrugation between the eyebrows. There may be excessive secretion of the lacrimal glands. According to Haig, the salivary and renal secretion may be diminished during an attack of migraine.

LABORATORY FINDINGS.—Bioglio found in studying the urine that nitrogen metabolism is slightly retarded in migraine during the interval between attacks; the amount of chlorids, total sulphuric acid, and earthy phosphates is below normal. Elimination of phosphoric acid is normal. During the attack the nitrogen elimination is increased; the other elements may or may not vary. He found a marked difference between the epileptic and the hemicranic with regard to metabolism.

Sicard and Cambessedes found that in simple or ophthalmic migraine the cerebrospinal fluid may be normal, or show an increase in albumin with or without hypercytosis. They report a case of ophthalmoplegic migraine in which the cerebrospinal fluid was normal.

Diagnosis.—Much confusion has been produced in the literature by using the term *migraine* when the term *headache* should be employed. Thus cases of sinusitis or eye-strain producing a one-sided headache are quite commonly termed migraine. It should be remembered that headache is a symptom and migraine is a disease. Migraine, periodic sick headaches, and bilious headaches, are terms used to designate recurring periodic attacks, in which headache is the chief symptom, which occur in definite attacks, the patient being in good health between them.

Headache is a symptom of many body disorders which produce this symptom whenever there is an unusual strain or acute exacerbation of the trouble. Either migraine or headache may be unilateral or bilateral; the situation of the pain cannot be accepted as the point of differentiation. In headache, the pain is apt to be less severe than in migraine; it is usually more prolonged, is relieved by the relief of the cause, such as eye-strain, sinusitis, constipation, fever, etc., and does not follow an orderly sequence of events as in a typical attack of migraine, such as prodromata, an aura, headache, vomiting, sleep, and relief, without apparent immediate provocation; or apparent immediate cause of re-

hef The family history of heredity, periodicity, good health between attacks, duration of the attacks, are important points in diagnosis.

Migraine must be distinguished from *cerebral tumor*, *aneurism* of the cerebral arteries, and *uremia*. Migraine is sometimes a premonitory symptom of *tabes* or general paralysis of the insane. It may disappear after the *tabes* develops.

Children between the ages of 2 and 12 years are often affected with *cyclic vomiting*, girls more often than boys; this tends to be replaced in adult life by migraine.

Migraine must be distinguished from *epilepsy* in rare instances. Usually there is no question of diagnosis but attacks may occur which are very similar and the diseases sometimes coexist, or one may entirely replace the other. The chief differences are: the *aura* in migraine is often visual; rarely so, in epilepsy. The visual aura of migraine consists of dimness of vision, or blotchy, irregular areas of darkness, which are one-sided, and prolonged, lasting from five to twenty-five minutes, or may consist of scintillating scotomata, white or colored lights; while in epilepsy, there may be only a single momentary flash of light, bright spots, or the momentary vision of a face, animals, a landscape, or other complicated visual impressions, lasting only a second or a few seconds. In epilepsy the aura is more often epigastric than in any other one location. In other cases of migraine, a sensation starts in a hand and gradually ascends the arm, leaving numbness behind it, and may ascend to the lips, but not to the head. In migraine, or epilepsy, the attack may be aborted at this point, but in some attacks the migraine will go on to the development of its characteristic feature, headache, while in epilepsy some attacks will go on to the production of a convulsion or the loss of consciousness; the occurrence of the aura without the characteristic climax in a given patient can be interpreted in the light of other complete attacks in that patient. The loss or clouding of consciousness in epilepsy is in contrast to migraine. The vomiting in migraine usually occurs after several hours of headache, but it may exist alone as an equivalent of a migraine attack, or precede the migraine by some years, as attacks of periodic vomiting. In epilepsy, the vomiting occurs during the fit. The characteristic feature of migraine is headache; this lasts hours or days. The characteristic feature of epilepsy is loss of consciousness or convulsions; these last usually only seconds or minutes (three to fifteen minutes). The headache is no essential part of an epileptic attack; when it occurs, it follows the sleep. The sleep following epilepsy is deep, profound, approaching coma, while that of migraine is less profound.

According to Forli, migraine and epilepsy develop on a neuropathic basis, but have a separate genesis and a different evolution.

Complications and Sequelæ.—The arterial spasm which occurs in migraine, if prolonged, may give rise to thrombosis or encephalomalacia. Among the complications of migraine are *paralyses* of the extrinsic or intrinsic ocular muscles (*ophthalmoplegic migraine*). The nerve most frequently affected is the third nerve; this may involve only the ex-

trinsic branches, or the whole nerve may be involved; in other cases only the intrinsic ocular muscles (iris and ciliary muscles) are affected (Bouchard). In all cases of paralysis of ocular nerves, the paralysis is on the same side as the pain. In one of Hunt's cases, there was an attack of migraine with complete paralysis of the left third nerve and a year later another attack followed by incomplete palsy of the right third nerve. Sometimes the sixth nerve alone is affected, and Hunt reports two sisters who had migraine and isolated abducens palsy developing during the attacks. One had left abducens paralysis during an attack of left migraine, while the other had a right abducens paralysis during a right migraine (though her migraine was usually on the left side). Flatau has collected 97 cases of ophthalmoplegic migraine. According to Hunt, isolated palsy of the trochlearis muscle following migraine has been reported by Luzenberger, and also by Bornstein.

Sometimes *hemianopsia* develops. This is usually homonymous. Hunt has reported a case of unilateral retrobulbar neuritis with paracentral scotoma, and Oppenheim a case of optic neuritis with paracentral scotoma. Unilateral optic atrophy, due to hemorrhage in the optic sheath or thrombosis of the central artery of the retina, has been reported. Hemorrhages into the retina and detachment of the retinal membrane have been observed during migraine attacks.

Aphasia occurs in some cases of left-sided migraine. It is usually temporary, rarely a permanent effect. *Organic hemiplegia* sometimes follows migraine attacks, and permanent *paresthesia* and *athetosis* sometimes occur.

According to Hunt, a recurrent facial paralysis has also been ascribed to migraine ("facioplegic migraine"); but both this case, reported by Rossolimo, and the case of transient *hypoglossal nerve paralysis*, reported by Sil, are isolated occurrences of inconclusive value. In Hoeflmayr's case there was spasm of one eyebrow and eyeball, and in one of the author's cases, permanent elevation of the left eyebrow in left-sided migraine.

Crouzon and Chatelen report a case in which in one attack the sixth nerve was paralyzed, and in four subsequent attacks the third nerve was paralyzed; in a later attack, the headache was omitted, but sudden complete paralysis of the third nerve occurred during the night.

Metz found records of 50 cases of herpes zoster ophthalmicus associated with ophthalmoplegia (third nerve in 32 cases, sixth nerve in 8 cases, fourth nerve in 4 cases, all in 5 cases). This is more significant, since Rohrer often found herpes labialis soon after attacks.

In some cases *unconsciousness* may occur (Hoeflmayr), or convulsions or tremor in rare instances.

Symptomatic migraine may occur in general paralysis of the insane, tabes, or brain tumor.

Association with Other Diseases.—Migraine is usually an independent hereditary disease. It is often associated with *xanthelasma*.

As *gout* and *rheumatism* often bear an etiological relation to migraine, they are, as would be expected, often associated in the same subject,

though often at different times. Jones found that rheumatoid arthritis patients may suffer for years from Raynaud's disease, asthma, or migraine before the joint swellings appear.

As a part of the same syndrome, vascular changes and *high arterial tension* often exist with migraine, and tend to develop earlier than in non-megriminous subjects. The occurrence of migraine in early life makes it improbable that the high arterial tension is the cause of the migraine.

Migraine may be associated with *neurasthenia*, *hysteria*, *writer's cramp*, *convulsive tic*, *brain tumor*; its association with *epilepsy* has been mentioned above.

Gowers maintains that migraine and epilepsy are closely related, as shown by their alternation, by the occurrence of one condition when the other is relieved, and by the occasional cases in which the same premonition may usher in either one or the other attack. In Kovalesky's case, the migraine was replaced later by epilepsy, and the attacks were preceded by an aura of a red light.

Hubbell says that he has seen 1,500 cases of migraine and none of them had epilepsy, or epileptic ascendants or descendants (?). Pappenheim reports a case of migraine which began in boyhood with auditory phenomena; later the patient had fainting attacks, still later melancholia, then tabes.

According to Oppenheim, *cardialgia*, violent *giddiness* or *convulsions*, may vicariously take the place of migraine. *Vomiting* attacks, *cyclical vomiting* or *periodic vomiting* of children may later be associated with migraine, or disappear when the migraine develops.

Hyperchlorhydria and *gastrosuccorrrhea* may be associated with migraine and are said to bear an etiological relation to it, although it is more probably a result.

Migraine and *angioneurotic edema* may coexist; or in other cases, other vasomotor disturbances, such as *coryza*, and unilateral sweating have been observed.

When *hysteria* is associated with migraine, hemianesthesia may occur on the same side as the pain (Oppenheim).

Ophthalmic migraine, when it appears first in the fourth or fifth decade, is often a premonitory symptom of *cerebral lues*, *tabes*, or *general paralysis of the insane*.

According to Forli and Oppenheim migraine may occur in cerebellar disease.

Hughlings Jackson has called attention to the close relationship between *chorea*, *migraine*, and *rheumatism*. Of 76 patients with chorea, 53 were subject to paroxysmal headache; in 31 of these the headaches were associated with nausea and vomiting, while in 14 ocular phenomena occurred. Migraine is more frequently associated with rheumatic chorea than with rheumatism without chorea, as it indicates cerebral involvement.

Clinical Varieties.—*Ophthalmoplegic migraine* is characterized by recurring attacks of migraine, beginning in early life usually, and eventually by associated paralysis of one or more ocular nerves recur-

ring in repeated attacks, the paralysis often becoming more marked and more permanent with succeeding attacks. The paralysis is on the same side as the pain, and usually upon the side of habitual attacks, although rarely the migraine and paralysis may both shift to the opposite side. The third nerve is most commonly affected; this may be the entire nerve, or only the extrinsic branches, or only the intrinsic branches may be affected. Rarely the fourth or sixth nerve may be affected, or one or both of them may be paralyzed in association with third nerve paralysis. When the third nerve alone is affected there is external strabismus, diplopia, mydriasis, with loss of reflexes to light and during convergence. At first this paralysis passes away entirely in a few days; later it may last several weeks or months, and eventually may become a permanent paralysis. After each attack of migraine the paralysis tends to last longer than the time before. Attacks of ophthalmoplegic migraine usually last two or three days, while ordinary migraine usually lasts one day.

In ophthalmoplegic migraine, the migraine almost always begins before 25 years of age, which is usually true also of ordinary migraine. Both occur in women twice as often as in men. Ophthalmoplegic migraine is invariably unilateral, but in a subsequent attack both the pain and paralysis may be on the opposite side. In ordinary migraine, it is unilateral in only 50 per cent. of cases. The pain always precedes the paralysis.

In *hemicrania sympathicotonica*, or *angiospastica*, there is mydriasis, widening of the palpebral fissure, hyperhidrosis.

In *hemicrania sympathicoparalytica* there is miosis, a narrow palpebral fissure, anhidrosis.

The symptoms of irritation and paralysis may be mingled.

Ophthalmic migraine is one of the most frequent types, in which the ophthalmic branch of the fifth nerve is affected. The attacks are often preceded by visual hallucinations, such as flashes of light, which may be white or colored, and may change in subsequent attacks. In one case the use of bromids changed the white lights into red. Sometimes there is scintillation or shimmering before the eyes (irritation); or there may be dark spots or fortification figures to one side of the fixing point (paralysis). The figures may be zigzag in shape and may be white, colored or black; or the hallucination may be more complex; and objects, landscapes, people or animals may be seen. There may be dimness of vision (amblyopia); or even temporary loss of sight (amaurosis); or blindness on one-half of the visual field (hemianopsia, which is, as far as the writer knows, always homonymous); or a paracentral blindspot (scotoma) may occur, which may be represented by bright lights (teichopsia) or darkness (amaurosis partialis fugax). Visual auræ are more common with ophthalmic migraine than with any other type.

Dysphrenia hemicrania transitoria is a term which has been applied to mental disturbances associated with migraine, such as transitory insanity, delusions, confusion, excitability or stupor, often associated with hallucinations, especially of sight.

Hemicrania cerebellaris was used by Oppenheim as a name for one case which he reported in which each attack of migraine commenced with typical cerebellar symptoms, difficulty in standing or walking, drunken gait and severe vertigo.

Hemicrania permanens, or *status hemicranicus*, may occur in people subject to migraine, in which more or less permanent pain is present.

Idiopathic migraine is the form in which no cause can be found, and these are usually the *hereditary familial type*.

Treatment.—DURING THE ATTACK.—Treatment during the attack is usually directed toward relieving the pain. The pain may be so severe as to require **morphin**, especially as the stomach often stops absorption and anything put into it may not be absorbed into the system during the pain, or may be vomited as soon as swallowed. The majority of patients are more comfortable lying down, and prefer a dark room with absolute quiet, and no food during the attack. Often emesis will bring the attack to an end, while in other cases a brisk **purge** affords relief. Hot or cold applications may be tried on the head, or rubbing with **camphomenthol**. **Blisters** applied at the point of pain are often helpful. In ophthalmic migraine they should be applied over the temple. **Setons** are rarely used now, and **leeches**, though valuable in plethoric cases, are almost forgotten. When the pain is in the eyeball, a drop or two of **cocain** in the eye will often give relief, while in cases of nasal obstruction a **cocain spray** in the nose may quickly terminate the attack. In most cases without morphin the patient serves out his allotted time and the attack comes to an end at the customary hour. Analgesics such as **acetanilid**, **acetyl salicylic acid**, **phenacetin**, **antipyrin**, **exalgin**, **pyramidon**, etc., are valuable in some cases and may be combined with **cafein**. The writer has found a combination of **acetanilid** (grains iii [0.2 gram]), **camphor monobromate** (grain i [0.065 gram]), **cafein** (alkaloid) (grain ss [0.0324 gram]) and **codein** (grain ss [0.0324 gram]) especially valuable. Hot coffee is often helpful during the attacks.

Aconitin grain 1/200 is recommended in ophthalmic migraine.

Inhalations of amyl nitrate or the administration of 10 to 20 drops of **chloroform** will occasionally abort an attack. **Tr. gelsemium** (gtt. viii) alone or with **Ext. cannabis indica** (grain 1/5 [0.13 gram]) are recommended by Wilcox. **Antifebrin**, **strychnin**, or **nux vomica** may help.

BETWEEN ATTACKS.—The cause should be sought for and removed if possible (*see* Etiology). In syphilitic cases, the usual treatment of **arsenic**, **mercury**, and **iodids** valuable. In cases that are not syphilitic but in which there is meningeal thickening on the base of the skull, **iodids** often do much good. The mode of life is important. Many of these cases find that over-work, studying, traveling, loss of sleep, excessive mental exertion, will bring on attacks, and these should be avoided. The bowels should be made to move well every day. Autointoxication is said to be a frequent cause and **alkaline waters** and daily **salines** are often helpful. **Gastric lavage** will sometimes help.

In rheumatic or gouty cases, the usual diet for these diseases should be followed. Some patients find that certain foods provoke an attack,

or think so, and these should be avoided. In these cases the suspected food will not produce an attack just after an attack has occurred, but will do so if taken after the usual interval between attacks has elapsed. In some cases diuretics are valuable, such as **lithium carbonate** or **diuretin**.

In malarial cases, **quinin** should be used.

In nervous cases, **bromids** are valuable and should be kept up for a long time. In Rohrer's case, **sodium bromid** changed the color of the scintillation to yellow, blue and red colors. Gowers recommends a combination of **nitroglycerin** and **strychnin**, given thrice daily for a long time to produce vasomotor stability. **Sodium nitrite** is used in cases where vascular dilatation is desired and **adrenalin** or **ergot** when vascular constriction is advisable.

The **eyes** should be examined in all cases, especially in regard to myopic or hypermetropic astigmatism, and muscular unbalance. The latter should be actually tested, and never guessed at. In Alexander's case, after wearing glasses, the patient continued to have premonitory symptoms but no pain, but the color changed to red and the quadrant hemianopsia to the right upper field. The author has seen cases who had been given glasses for astigmatism, etc., without relief; subsequently esophoria, exophoria or hyperphoria were found and the cases relieved. Often the muscular unbalance is not primary. It may be the result of the migraine (ophthalmoplegic migraine) or associated with it; or in other cases, it is due to anemia, which itself is due to some chronic infection, such as unsuspected tuberculosis, chronic tonsillitis, abscessed teeth, etc., and the primary condition and anemia need attention. **Iron** and **arsenic** are valuable in these cases.

Adenoids or hypertrophied tonsils should be removed. Chronic nasal disease preventing free drainage of the sinuses should be properly treated. Sometimes a **camphomenthol spray** suffices; in other cases an operation is indicated.

Climate.—A **change of climate** is beneficial. It probably does good by getting the patients away from work and worry and creating a cheerful, pleasant attitude in agreeable surroundings. It does not seem to matter whether they go to the mountains or seashore, as long as their change is a pleasant one. Sometimes the attacks cease after a change of climate.

Physical Agents.—**Baths** are often helpful especially in gouty or rheumatic, or toxic cases. Cabinet baths followed by alternating hot and cold **douches** to the spine (Scotch douches) are helpful. **Massage** helps the general condition of the patient. **Electricity** is probably of more psychic than direct value. Daily **exercise**, especially agreeable games out of doors, is valuable.

Organotherapy.—In cases of hypothyroidism Sapogenix recommends the use of **thyroid gland** (grain ss [.0324 gram] t.i.d.) in ophthalmic migraine. She obtained favorable results in 10 out of 23 cases.

Diet.—Osler recommends avoidance of excitement, **regularity** in meals, moderation in **diet**, and especially a vegetable diet.

Prognosis.—Typical migraine begins usually in childhood or early life, nearly always before the age of twenty-five—rarely later. It is hereditary and familial, and usually lasts more or less a lifetime. Nevertheless, it may cease spontaneously at any time; or in cases where a cause is found, it may be relieved by proper treatment. Even in idiopathic cases persistent treatment sometimes gives relief but must be kept up for several years. There is a tendency for the disease to cease at the menopause, but not always. While the disease sometimes begins with the onset of the menses or with pregnancy, it has also been known to cease at these times. The disease usually ceases in old age, especially after fifty years of age; in cases that persist, the attacks become milder.

The danger to life is not great, but some of the cases develop thrombosis of the cerebral vessels and die from this cause.

In a few of Oppenheim's cases the migraine disappeared after an injury to the head, and one case ceased for twelve years after typhoid fever.

The danger of insanity is not great. When parents are insane and one of the offspring has migraine, the latter is less liable to insanity than the other children.

Permanent visual disturbance may follow the transient visual phenomena of migraine. Ophthalmic migraine may be followed by permanent aphasia, ophthalmoplegia, hemiplegia, hemianopsia, or amaurosis. In the majority of attacks, however, these symptoms pass off, but may recur.

Migraine may disappear for years and then recur.

The vomiting that occurs in childhood with migraine may cease entirely as the patient gets older. Hysterical migraine offers a fairly good prognosis. When the migraine affects first one side and then the other (irregularly), the prognosis is better than when it is always confined to the same side.

According to Clarke, the aura, aphasia or hemiopia, may become permanent; in others the permanent disability in no way corresponds to the aura.

Marriage or childbirth may cause a cessation of the attacks. They often get worse near the menopause.

Pathology.—The pathology of idiopathic migraine is not known. In fatal cases thrombosis of cerebral vessels, such as the carotid, with softening due to vascular disease such as endarteritis, has been reported. Sometimes hemorrhage occurs; but it is a question whether the vascular changes are cause or effect. They are certainly not the cause of the pain of migraine, which begins so often in childhood. Thomas concludes that while in the majority of cases described, vascular or cerebral disease existed, and the migraine was symptomatic, there is reason to believe that, as a result of idiopathic migraine, vascular lesions may occur in young persons with healthy vessels.

According to Knapp, in autopsies on cases of ophthalmoplegic migraine, the lesion has not been of a vascular character. There were either meningitic disturbances, or a tumor in the neighborhood of the

third nerve. Hunt cites several such cases: fibrosarcoma attached to the third nerve (Thomsen-Richter); neurofibroma of the dura pressing on the nerve (Karplus); granuloma nodule on the nerve (Weiss).

Déjerine says that ophthalmoplegic migraine is generally produced by tumors or meningeal exudates of syphilitic or tuberculous nature affecting the base of the skull. In two of the author's cases x-ray photographs showed distinct thickening on the migraine side of the sella turcica, with paralysis of the oculomotor nerve in both cases. A third case showed nothing abnormal in the x-ray picture.

Gübler (quoted by Fisher) has described a plastic exudation in the basal subarachnoid space in one case, in which fibrous adhesions were also present around the origin of the third nerve. In other cases a fibroma or chondrofibroma involving the ocular nerves has been described. No nuclear lesions have ever been noted.

Spitzer suggested that it may be caused by abnormal narrowness of the foramen of Monro, so that, when hyperemia of the choroid plexus occurred, the ventricular fluid caused distension of the lateral ventricles and pressure of the hemispheres against the skull cap.

The chief theory of migraine is that the attacks are of vasomotor origin, and that, whatever else may provoke it, the actual attack is due to vasomotor spasm or dilatation. According to Brunton, the proximal ends of the arteries are dilated, while the distal ends are constricted in migraine. As the caliber of the vessels is regulated by the sympathetic nerves, we assume that migraine is of nervous origin. As a result of these vascular disturbances there may be arterial changes of a permanent character, such as thickening of the walls of the temporal artery on the side of the migraine, sometimes hemorrhage from the diseased blood-vessels in the brain or eyeball, or sheath of the optic nerve, or thrombosis from prolonged stasis in the spasmodically constricted artery; as a result of this, encephalomalacia may occur, producing symptoms and tract degenerations in the brain and cord, depending upon the seat of the lesions.

In Bouchard's case of ophthalmoplegia interna, he attributed the paralysis to spasm of the arterioles supplying the third nucleus, followed by narrowing of the vessels producing malnutrition (ischemia) of some of the nerve cells. Thomas quotes two cases of Galizowski's of permanent hemianopsia from arterial thrombosis or embolism in young subjects occurring in migraine attacks.

As to the seat of the lesion in migraine, it seems probable that it varies, but is most often in the cerebral cortex. The occurrence of psychical symptoms, aphasia, agraphia, astereognosis, are suggestive of a cortical seat. Furthermore, the relation of the symptoms to each other suggest a cortical origin. In left-sided migraine we may have numbness of the right arm and right homonymous hemianopsia; or a combination of aphasia with left migraine and right hemiplegia; migraine with right hemiplegia; right astereognosis, and motor aphasia (transient attack); aphasia and right hemiplegia; left hemiplegia with left hemianopsia; right hemiplegia with athetosis in the right hand; right mi-

graine and right paracentral scotoma; numbness in the right hand with right homonymous hemianopsia.

The absence of the hemiopic pupillary reflex in one of Hunt's cases, with the absence of darkness in the blind half of the visual field, point to the lesion being back of the primary optic centers. On the other hand, he reports a case of left migraine with numbness in the right hand, and *blurring* of the *left* field of vision, with a subsequent attack accompanied by persistent left homonymous hemianopsia.

Eldridge-Green contends that the condition present in migraine is a central scotoma increasing from within outward. He states that any disturbance of the circulation in the eye preventing the flow of photochemical fluid to the fovea would produce a central scotoma increasing from within outward.

Levi thinks the disease arises from the medulla oblongata. It would be very easy to account for the chain of symptoms on this supposition, such as the nausea (ninth nerve), vomiting (tenth nerve), vertigo (Deiters' nucleus), hemiplegia (pyramidal tracts), etc.; but this offers no explanation for the hemianopsia, aphasia, psychic disturbances, etc.

Alexander quotes Noyes as recording a case of persistent hemianopsia following attacks, with softening of the cuneus found at autopsy.

It seems most probable that the symptoms of migraine are produced by a localized vasomotor spasm of the vessels of the cerebral cortex, and that the pain is of peripheral nerve origin.

"Observations made by Monglesdorf show that in every case of migraine there occurs a well-marked, acute dilatation of the stomach, and that a frequent repetition of these dilatations leads to a permanent gastric atony. Monglesdorf claims to find these same dilatations in epileptics during the attacks, which would be another point in evidence of close resemblance between epilepsy and migraine" (Kelly).

Historical Note.—The history of migraine can be obtained in Campbell's book. The first case of ophthalmoplegic migraine was described by Gübler in 1860. Möbius reported recurrent "periodic ocular paralysis," but Charcot gave the disease a distinct clinical entity and applied the name "migraine ophthalmoplegique."

Sociological Aspect.—If people with migraine wish to marry, they should consider the possibility of the transmission of the disease to their offspring. The marriage of a man with migraine is far less serious than the marriage of a woman, as the disease is transmitted usually through the female line. A man with migraine may have a daughter free from it, but her children, whether male or female, may be affected (atavism). In a general way, it may be said that the inadvisability of marriage is proportionate to the number of cases in the family, the number of generations affected, and the severity and frequency of the attacks (*see* Prognosis).

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CHAPTER XX

EPILEPSY

By E. BATES BLOCK, M.D.

Definition, p. 401—Etiology, p. 401—Symptomatology, p. 414—Diagnosis, p. 428—Differential diagnosis, p. 429—Complications, p. 434—Sequelæ, p. 434—Association with other diseases, p. 435—Clinical varieties, p. 436—Treatment, p. 438—Prophylaxis, p. 438—General management, p. 438—Diet, p. 439—Treatment of causes, p. 439—Treatment during the attack, p. 440—Treatment of status epilepticus, p. 441—Treatment between attacks, p. 441—Bromids, p. 441—Other drugs and methods of treatment, p. 444—Surgical treatment, p. 447—Prognosis, p. 448—Pathology, p. 452—History, p. 455—Distribution, p. 455—Sociological aspects, p. 456—Bibliography, 457.

Synonyms.—Falling sickness, morbus sacer, spasms, fits, spells.

Definition.—No definition of epilepsy can be given which will include every case of epilepsy and at the same time exclude every other disease. As a fair proposition, however, we may say that *epilepsy is a disease in which there occur repeated transient attacks of either a psychic, sensory, or motor nature with a loss or impairment of consciousness.* The only objections to this definition are that it does not include the first attack and does not include every case of jacksonian epilepsy. However, a diagnosis of epilepsy can never be made on one attack, and jacksonian epilepsy is a special chapter in itself.

The essential feature of epilepsy is a repeated and sudden loss of consciousness with or without a convulsion, and without apparent immediate cause.

Etiology.—Anybody who is looking for one cause of epilepsy might as well stop looking. Epilepsy is a symptom of many disorders—organic, physiological and chemical. It seems evident that epilepsy is not due to one cause, but to a combination of causes, which are necessary to produce the attacks. Certainly, while dentition, worms, adhesions of the prepuce, etc., often cause convulsions, the majority of the children having these conditions do not have convulsions. The other factor, therefore, must be looked for in the brain and the only defect which seems apparent is cerebral instability. This has been termed “*spasmophilia*” by Féré, or the “*convulsive aptitude*” by Joffroy. In support of this view it may be stated that about 25 per cent. of mentally defective children have epilepsy, that we rarely find epilepsy in the children of strong, healthy parents except in cases that have had some cerebral trauma, such as accidents, hemorrhage, forceps delivery, meningitis, etc. The improvement in mental stability

(as in the treatment of cretinism or removal of adenoids, etc.) is followed by improvement in the convulsions.

According to Turner, "it is a cardinal principle that the cause of epilepsy is that circumstance to which the first fit is apparently due." After that the spells occur independently of any obvious cause.

As to the seat of the disease, there have been various opinions held—the medulla, the pons varolii, the subcortical centers—but it seems evident that the seat of the motor discharge is in the highest cerebral cortical centers, either the psychic, sensory or motor—or even, more probably, the psychosensory, psychomotor, psychovisual, etc., centers. In favor of this is the loss of consciousness, the psychic attacks, the clonic nature of the fits, the production of attacks by brain injury, the mental state of epileptics, etc. We must not lose sight of the initial loss of consciousness and petit mal.

IMMEDIATE CAUSE OF THE ATTACK.—Jackson thought the attacks due to a sudden violent discharge from a nerve center, psychic, sensory or motor, due to congestion and excess of blood in the cortical capillaries with retarded blood motion and consequent alteration of the nutrition of the cortical cells. Following the attack, there is an exhaustion of the nerve-cells until food is again stored in them to produce energy for another attack, much like a Leyden jar.

According to Turner, the onset of a fit may be accompanied by temporary cessation of the heart's action and the pulse beat, due to cardiac inhibition, and the loss of consciousness and the fit are attributed to a sudden cerebral anemia. The convulsions in Stokes-Adams' syndrome and the Kussmaul-Tenner fits from ligation of the internal carotid arteries, spasms from stimulation of the peripheral end of a cut vagus nerve, are attributed to cerebral anemia.

A. E. Russell considers the fundamental factor underlying the epileptic fit to be cerebral anemia. Undoubtedly there exists in epilepsy a vasomotor instability. Russell thinks that the unconsciousness and the fit are the results of cardiac inhibition. He quotes Fox as attributing the loss of consciousness and tonic spasm to cerebral vasoconstriction, and the clonic convulsion to gradual yielding of the vasomotor constriction allowing some, but not enough, blood to reach the brain for complete cerebral control. In support of Russell's opinion, the retinal arteries have been found narrowed on ophthalmoscopic examination during the paroxysmal attacks of impairment of vision which occur in some cases of epilepsy. Gowers did not observe any constriction of the retinal arteries at the beginning of an epileptic attack. If the heart ceases to beat just before epileptic fits it is probably due to the influence of the brain on the heart, rather than the influence of the heart on the brain. The headache following the fit is probably due to venous engorgement of the cerebral membranes.

Cushing has shown that a rapid increase in intracranial pressure causes "Kussmaul-Tenner spasms, evacuations of the bladder and rectum, practical cessation of respiration, and pronounced vagus effect upon the heart, often with a complete standstill lasting from 10 to 20 seconds."

Gibson, Good and Penny believed that, in their two cases in which they were obtaining pulse tracings at the time of the fits, there was no lowering of general blood pressure sufficiently marked to account for the fit on the assumption of cerebral anemia due to a general cause.

Turner calls attention to the fact that paralysis of the vasoconstrictor nerves from removal of the superior cervical sympathetic ganglion does not cure epilepsy.

Berger does not think epileptic attacks are brought about by cerebral anemia, and Osler expresses the same view. In cases examined by the writer during the fit, while the pulse felt feeble, the heart beat rapidly and forcibly during the attack. The apparent congestion of the face is due to venous stasis from cessation of respiration.

Turner gives reasons in support of the thrombotic origin of epileptic fits. (*See Pathology.*)

Starr gives the following table of alleged causes of epilepsy:

Trauma	229	Sunstroke	12
Fright	119	Physical strain	11
Alcoholism	61	Measles	12
Invaliddism	49	Indigestion	10
Mental strain	36	Arteriosclerosis	8
Menstrual disorders	24	Syphilis of brain.....	8
Dentition	23	Whooping-cough	6
Childbirth	24	Rickets	5
Scarlet fever	20	Excessive heat	5
Infantile palsy	19	Trauma to mother.....	4
Maldevelopment of brain....	19	Diphtheria	4
Masturbation	14	Typhoid fever	4
Menopause	11		

The above causes were given by the patients or their families as their opinion. In 1,363 cases out of 2,000 no such cause could be ascertained.

According to Moon, in 50 per cent. of cases of convulsions in children, the first convulsion arose without any obvious cause.

OCCURRENCE.—Frequency of Spells.—As a rule, we may say that patients afflicted with minor epilepsy have a far greater number of spells than those afflicted with major spells. Also as a rule the patients with major spells have them more frequently if they are mild than if severe. The more severe, the further apart are they apt to occur. The frequency of attacks varies greatly in different subjects. They may average one a year, or one a month, one a week, or from one to one hundred in twenty-four hours. Generally a patient can tell approximately how many to expect in a given period of time, but in some cases they appear in crops so that a patient will have a series of attacks during one day or one week or month, and then have an interval of freedom from them till the next series starts (serial epilepsy). Quite contrary to expectations, Thom and Southard found that cases with apparently normal brains had convulsions more frequently (one or more a day) than cases with abnormal brains (one or less a month).

Spratling reports a case in which a woman had 519 attacks in 49 hours, and a case of petit mal who had 26,124 spells in 5 years without impairment of her mental faculties.

The frequency of the attacks is well shown by the following table taken from Spratling:

Attacks weekly or oftener.....	57 per cent.
Attacks every 2 weeks	13 per cent.
Attacks every 3 weeks	4 per cent.
Attacks every 4 weeks	18 per cent.
Attacks every 8 weeks	3 per cent.
Attacks every 12 weeks	4 per cent.
Attacks every 6 months or over	2 per cent.

Attacks are said to be more frequent on a new moon than on a full moon (Michalek).

Frequency of the Disease.—Spratling gives the following interesting figures comparing the number of epileptics to the population in various countries:

In Switzerland.....	1 to 750 (Kolle)
In Russia.....	1 to 2,000 (Kovalevsky)
In Russia.....	1 to 1,000 (Shoutelwort)
In Scotland.....	1 to 750 (Pelmann)
In France.....	1 to 1,100 (Vernet)
In United States.....	1 to 500 (Peterson)

There are over 150,000 epileptics in the United States.

Periodicity.—There is often a remarkable periodicity in the spells. One of the writer's patients had only five spells, which occurred every June for five successive years, then ceased under treatment. Another case had one spell at the end of each month (male patient), so that he could predict practically to the day the onset of his attacks. Sometimes the periodicity is interrupted, either from treatment or the influence of some other unknown factor, by the elision of a period, to be resumed at the next time.

Season.—There is no season of the year when spells do not occur, but they are apparently more frequent in the spring or when a warm spell follows cold weather.

Time.—The attacks may occur at any time, day or night. In the author's experience they are more frequent in the early morning hours before or on waking. They are less frequent during sleep, but may occur either while the patient is asleep or awake. When they occur only during sleep they may occur either at night or in sleep in the daytime. Whatever time of day or night a patient has the spells, he can usually count upon future ones coming at the same time, although they may come at any time. Thus one of the writer's patients had all of her attacks at 6 A.M., except one which came at midnight; another patient has

had all of her attacks between 2 and 4 P.M. Another patient has all of his attacks between 7 and 10 A.M.

Turner says the frequency of attacks an hour or two after falling to sleep has led to various theories as to cause, namely, brain anemia (Pick), lessened inhibitory control (Clark), the alkaline tide (Haig), increase in the rate of blood coagulation (Turner).

According to Spratling, in men the attacks are of equal frequency in the day time and at night time, while in women five-eighths occur in the day and three-eighths in the night. In a summary of over 100,000 attacks, Spratling found practically no difference in the frequency of attacks in the day and night; the largest numbers occurred, however, at 2 and 3 A.M., and the smallest number at 7 P.M. In the author's cases, the early morning hours, before breakfast, show the greatest frequency.

Sex.—Males are slightly more often affected than females according to Osler, Althous, Spratling and others, while Gowers found 53.4 per cent. females and 46.6 per cent. males. Spratling states there are 5 men to 4 women.

Age.—The disease may occur at any time up to death in old age. The earliest cases are often due to cerebral hemorrhage or cortical agenesis, forceps injury at birth or early trauma to the head. In childhood we should look especially for trauma, impacted teeth, adhesions of the prepuce, masturbation, worms, constipation, errors in diet, or syphilis. After 30 years of age we should think particularly of alcoholism, syphilis, dementia paralytica, brain tumor and cysticercus. In old age we should think of arteriosclerosis, chronic nephritis, senile involution, apoplexy, etc.

Gowers gives the following approximate figures of the age of onset of epilepsy:

0-10 years of age.....	25	per cent.
10-20 " " ".....	50	per cent.
20-30 " " ".....	14	per cent.
30-40 " " ".....	16.5	per cent.
40-50 " " ".....	2.5	per cent.
50-60 " " ".....	1.0	per cent.
Over 60 " " ".....	1.0	per cent.

Seventy-four per cent. begin before 20 years of age. Decidedly a larger number begin in the first three years of life (13.5 per cent.) than in any other similar period of life.

There is another increase in frequency at 7 years of age, at the time of the second dentition, and another rise in frequency at puberty. In Spratling's table the largest number is in the first year of life. He reports 82 per cent. occurring before 20 years of age. The usual statement is that one-third occur before 10, three-fourths before 20 years. One-eighth of the fits of infancy are due to epilepsy. The average age of onset for boys is 14, and for girls 16. According to Osler more cases began in the first or in the second year of life than in any other one

year, with a distinct increase in the fifteenth year as compared with ages from the fifth to fourteenth years.

IDIOPATHIC EPILEPSY.—The causes of idiopathic epilepsy are not evident on examination. Sometimes these cases are due to jars or jolts to the nervous system, concussion, or to small adhesions of the membranes, to small cerebral hemorrhages or localized encephalitis, none of which gives definite evidences at the time of examination on account of the development of epilepsy. It is probable that heredity plays a very important part in this class of cases. Idiopathic epilepsy is rapidly being reduced in frequency by more careful study of the cases.

Bolten regards idiopathic epilepsy as due to chronic autointoxication from alimentary decomposition or toxins of cellular metabolism which are insufficiently neutralized by the thyroid and parathyroid glands, and he finds that all symptoms disappear after rectal administration of fresh extracts of these glands. Such cases can no longer be regarded as idiopathic, however, as the term is now used to represent cases in which no cause is known.

EMOTIONAL DISTURBANCES.—Frights, excitement, anxiety, shock, grief, anger, coitus, are said by Gowers and others to be causes of epilepsy; in Gowers' cases they caused one-third of those in which a cause was given, and one-seventh of his whole series. The writer never saw a case in which he was certain that fright was the cause and it seems doubtful if emotional disturbances ever cause epilepsy, although it is a frequent cause of hysterical convulsions. Overwork and prolonged anxiety may, however, so undermine the nervous constitution as to allow epilepsy to take place in those predisposed to it. Starr and others think fright one cause of epilepsy.

Turner says that emotional disturbances produce epilepsy twice as commonly in girls as in boys.

Spratling found 5.5 per cent. of cases due to emotional shock, occurring more frequently in females than males, while he found that anxiety, grief and overwork are causes of epilepsy in 3 per cent. of females and 0.6 per cent. of males.

HEREDITY.—It has been said that, if either parent be drunk at the time of conception, it predisposes the offspring to epilepsy. Spratling found a history of *alcoholism* in 16 per cent. of the fathers and 12 per cent. of the mothers of epileptics.

Echeverria (quoted by Osler) out of 572 cases found 257 were directly due to alcohol, 126 in which there were associated conditions, such as syphilis and traumatism, 189 in which the alcoholism was probably the result of epilepsy.

Spratling found *tuberculosis* existed in 15 per cent. of the fathers and 12 per cent. of the mothers of epileptics, while others have found tuberculosis to be the most common disease in the ascendants and family of epileptics.

Chronic lead poisoning in the parent sometimes produces epilepsy in the offspring.

There can be no doubt that heredity plays a very important rôle in

the production of epilepsy. Southard very wisely remarks that the influence of heredity is permissive rather than mandatory. According to Spratling "inherited causes play their most important rôle in early life." He found direct heredity (similar heredity) in 15 per cent. of men affected and in 17 per cent. of women. Hereditary influences were present in 35 per cent. of Gowers' cases and 56 per cent. of Spratling's, when both similar and dissimilar heredity were classed together. The inheritance is from the mother's side more frequently than from the father's (6 per cent. difference), and women inherit epileptic tendencies a little more frequently than men. Osler found only 5 out of 435 cases were children of epileptic parents, in each case on the mother's side.

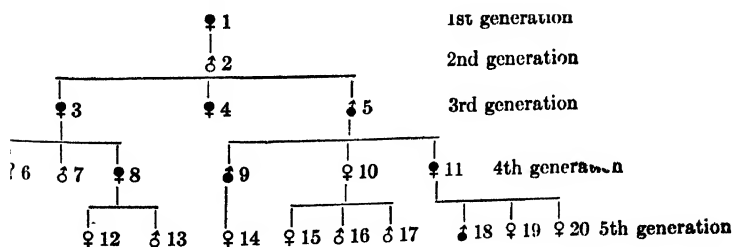
Among the diseases which furnish hereditary equivalents in the parents are hysteria, neurasthenia, migraine, insanity, obsessions, tics and many other functional diseases, and even organic diseases such as brain tumor. Oppenheim finds hereditary influence in from one-third to one-half of the cases, and quotes Binswanger 35 to 40 per cent., and in nearly three-fourths according to Finckh.

Spratling found that insanity was present in 7 per cent. of the fathers and 10 per cent. of the mothers.

Travelyan reported epilepsy in two twin Jewish boys with eight siblings, all of whom had fits while teething.

Clark thinks that a combined hereditary and toxic action are responsible for most cases of epilepsy. Stigmata of degeneration are very common in epileptics, showing an abnormal development. It is rare for idiopathic epilepsy to develop after puberty.

The influence of heredity is clearly shown in the family tree of two of the writer's patients. Of these cases, Numbers 7 and 8 were twins. Number 4 had convulsions at 70 years of age. In 5 generations 8 out of 20 in the family had convulsions. The Wassermann test was positive in the two cases under the writer's care.



The table of hereditary influence shown on the following page, representing the experience in France, Germany, America and England, is taken from Turner's article.

Mentally defective children are particularly liable to epilepsy (25 per cent.), and it may also be said that repeated epileptic seizures lead to mental enfeeblement.

TABLE SHOWING INFLUENCE OF HEREDITY IN EPILEPSY

	Déjerine	Binswanger	Spratling	Turner
Epilepsy.....	21.2%	11.0%	18.0%	37.4%
Insanity.....	16.8%	29.6%	7.0%	4.8%
Alcoholism.....	51.5%	22.0%	14.0%	4.0%
No heredity.....	47.0%
Totals.....	350 cases	150 cases	1,070 cases	890 cases

Stigmata of degeneration are very common in epileptics; malformations of the skull, abnormal ears, facial asymmetry, errors of refraction, malformations of the extremities, high arched palate, dental anomalies, etc., are very common. Physiological stigmata occur as well as anatomical stigmata.

ORGANIC BRAIN DISEASES.—Spratling found that 11 per cent. of epileptics at the Craig colony had had infantile cerebral palsies.

According to Osler, in more than 50 per cent. of the cases of infantile hemiplegia the affection (hemiplegia) follows severe convulsions. Some of the cases of infantile cerebral palsies with epilepsy are due to apoplexy, some to acute encephalitis, while others are due to cortical agenesis. According to Clark the heredity factor in hemiplegic epilepsy is only a little less in evidence than that present in genuine epilepsy.

Falls are frequently given as the cause of epilepsy, and it is not improbable that the general shaking up of the brain, or small hemorrhages produced by concussion and giving rise to scar tissue, or organization with adhesions of the meninges often produce the disease.

Injuries to the head may give rise to jacksonian epilepsy or to general convulsions of a non-focal character. Even with distinct focal evidences sometimes no lesion is found to account for the spells. Trephining is sometimes followed by convulsions, though rarely, and it is surprising how seldom this occurs when all the possibilities are considered.

The convulsions occurring in brain tumor, abscess, fractures, meningitis, multiple sclerosis, general paralysis of the insane, gummata, cerebral syphilis, etc., should be regarded more properly as symptoms of those diseases.

It is interesting to note that in jacksonian epilepsy the convulsions may cease after an apoplexy on the side of the brain lesion, or in bilateral epilepsy the convulsions may cease on the side opposite to the cerebral lesion. Thom and Southard found head injuries in 19.1 per cent. of their cases. Trauma is a frequent cause of epilepsy. Most often the injury is to the head and usually with a history of a loss of consciousness at the time of the accident, rarely with no history of unconsciousness. The writer was impressed with the frequency of scars on the scalp or head in cases of epilepsy when no history of an accident was given, and they can never be excluded without shaving the head. Injuries to the head may consist of concussion, hemorrhage, depressed fractures or bullets in the brain. Injuries to the spinal cord and peripheral nerves are said to be causes. Spratling found trauma to be the cause of epilepsy in about 3.5 per cent. of women and in 8.5 per cent. of men. The influence of trauma is not confined to head injuries,

but injuries to any part of the body can produce an impression on the brain which places it in a state of disequilibrium. Thus injuries to the hand or foot, with or without a scar, may be followed by epilepsy. The scar may be sensitive and irritation of it by pressure or a blow may cause an attack. Such lesions are spoken of as an epileptogenic zone. According to Oppenheim a concussion or traction on a limb may produce an irritation of the corresponding cortical center which thus acquires an epileptogenous condition. The epilepsy may develop many years after the injury which produces the scar, and the relation of cause and effect may be overlooked for this reason.

CIRCULATORY DISTURBANCES.—Atherosclerosis is a common cause of epilepsy in *epilepsia tarda* or senile epilepsy. The cerebral arteriosclerosis gives rise to disturbance of the circulation and malnutrition of the brain.

Heart disease, especially aortic stenosis, sometimes gives rise to convulsions, probably from malnutrition of the brain. Degeneration of the heart muscle and Stokes-Adams' disease probably act in the same way. Heart disease, while not necessarily the cause of epilepsy, is at least frequently associated with it, occurring in 238 out of 1,070 of Spratling's cases. Aortic aneurysm occasionally causes convulsions.

RESPIRATORY DISTURBANCES.—Laquer reports a case of an epileptiform fit following sudden asphyxiation from miswallowing a piece of meat.

TOXIC CAUSES.—*Auto-intoxication* has been considered by a great number of writers as the cause of epilepsy, some of them thinking it the most frequent cause, while some even go so far as to think all cases due to this cause, which, of course, cannot be maintained. Nevertheless, indican and skatol are often found in the urine, constipation is frequent, after the attack the urine is more toxic than normal, while between attacks it is less toxic. This last, however, may be due to the excessive body activity during the convulsion. Uric acid or ammonium carbonate have been regarded as the cause, also cholin (Donath and Justiss) and the organic ammonia bases (trimethylamin).

It may be said that the severer the attacks, as *status epilepticus*, serial epilepsy and major attacks, the greater are the amounts of poisons found in the blood and urine, which would seem to indicate that they are the result and not the cause of the convulsions.

Krainsky attributed the convulsions of epilepsy to the formation of carbamic acid, one of the derivatives of urea.

Of the substances found in the blood and urine of epileptics, Donath (quoted from Turner) did not produce convulsions in guinea pigs and dogs with uric acid, neutral urate of sodium, carbonate of ammonium, kreatin or lactic acid. While convulsions were produced by ammonia, and organic ammonium bases, trimethylamin, cholin and creatinin.

The use of the x-ray has given valuable information in reference to lesions of the colon. Kinks in the colon, malpositions, extensive dilations, adhesions, colonic stasis, have all seemed in some cases to be the cause of the seizures, while it cannot be claimed that it is the only

cause. Adhesions of the appendix and appendicitis belong to the same group of cases. One of the writer's cases developed epilepsy at the age of 38. The only pathological condition found was an enormous dilatation of the colon from using two gallons of water as an enema every morning for several years.

Overeating, overloading the stomach, or eating indigestible foods (bananas, peanuts, meats) is not an infrequent cause of convulsions in children and will produce convulsions in epileptic subjects. Sometimes there exist idiosyncrasies to certain foods, such as shellfish, and certain fruits, such as strawberries, which presumably produce an anaphylactic reaction similar to urticaria, etc., in the brain or meninges.

The convulsions occurring in alcoholic encephalitis, in lead encephalitis (saturnine epilepsy) and uremia should be regarded as part of the symptoms of those conditions rather than of epilepsy. Saturnine epilepsy occurred in only 1 out of 1,600 of Spratling's cases.

Intestinal putrefaction with the appearance of indican and skatol in the urine is certainly very common in epilepsy. Alexander found a relationship between a fit and a low urea-ammonia ratio. *Gout* is a doubtful cause of epilepsy. Tintemann in a case of diabetes with epilepsy found a relation between the amount of uric acid excreted and the number of attacks. Proteid sensitization probably plays a part in some cases of epilepsy, but the writer's observations with these tests have been inconclusive.

Convulsions are produced by many *poisons*, such as alcohol, lead, cocain, arsenic, chloroform, ether, physostigmin, antipyrin, strychnin, camphor, camphor monobromate, theophyllin, lumbar anesthesia by stovain. Marburg reported a case of epilepsy following many years' use of 25 to 180 grains (1.6 to 11.6 grams) of coffee a day.

ALCOHOLISM.—Kovalevsky found epilepsy commonest in the heaviest wine drinking district of Russia.

Chronic drinkers frequently develop epilepsy, especially whiskey and absinthe drinkers. "Wartmann found 206 drunkards among 452 male epileptics (Oppenheim), but half of these had been epileptic from their youth, while others were associated with other injurious circumstances."

EYE STRAIN.—In some of the writer's cases there were obvious defects about the eyes, muscular imbalance, astigmatism, hyperopia or myopia, with marked benefit from the fitting of proper glasses. According to Féré, astigmatism is present in 75 per cent. of epileptics.

REFLEX EPILEPSY.—According to Oppenheim "many cases show that injury of a peripheral, in particular of a cutaneous nerve, or its irritation by a cicatrix or a foreign body may produce spasmodic attacks of an epileptic nature." A cicatrix of the scalp and a lesion of the meninges may give rise to reflex epilepsy. Many facts seem to indicate that a stimulus arising from any part of the body may have this effect.

ADHERENT PREPUCE.—Adherent prepuce both in the male and female is a frequent cause of epilepsy. The condition is much more easily recognized in the male and is often not looked for in females. Frequently the attacks cease after the adhesions are pulled loose or after

circumcision, but more often sedative treatment is also necessary. The writer saw two cases of adherent prepuce in cases with a history of circumcision in which the adhesions had formed after the circumcision or were not pulled loose at the time of the operation. Phimosis has also been reported as a cause of epilepsy.

Nose.—Adenoids and nasal polypi are said to be causes of epilepsy and occasionally convulsions cease after their removal. Levenstein reports a case of a convulsion occurring immediately after the removal of nasal polypi, and quotes Lannois as reporting a similar case.

Diseases of the accessory sinuses of the nose, and laryngeal polypi have also been reported as causes.

Ears.—Foreign bodies in the ear and chronic otorrhea are said to be causes. Mendoza induced epileptic fits by pressure on an aural polypus.

Teeth.—Carious teeth and impacted teeth are said to be causes of reflex epilepsy, while apical abscess and pyorrhea have been suggested as possible sources of focal infection in some cases of late epilepsy by the author.

Dentition.—The occurrence of convulsions during dentition indicates a cerebral instability, but it depends upon the degree of this instability as to whether the convulsions will continue and constitute a true epilepsy. Certainly the frequency of convulsions during both the first and the second dentition shows that a relationship exists, although the incidence is less marked in the second dentition than in the first.

Menstruation.—Any apparent association with menstruation is probably due to the tendency to periodicity of the spells, although Oppenheim claims that they are often more frequent just before or during menstruation. One of the writer's patients who claimed a relationship between her attacks and menstruation did so because they came ten days after each menses, but such relationship is not claimed in male patients who have spells once a month, in one of the writer's cases almost regularly on the 28th of each month.

The menses are more frequent in chronic cases. Gordon reported 23 cases in whom the spells coincided with the menstrual periods. Toulouse and Marchaud say that the establishment of the menses can cause a disappearance of epilepsy or may aggravate it, and the same is true of the menopause. In some cases the beginning of the epilepsy seems connected with the appearance of the first menses. In the majority of cases the menses have no influence on the attacks.

Pregnancy.—The influence of pregnancy is uncertain. Cases have been reported in which there was a complete cessation of fits during pregnancy, while others have claimed that pregnancy sometimes causes epilepsy (question of eclampsia?). W. A. Turner states that pregnancy and the puerperium usually increase the attacks, or pregnancy may be the exciting cause of the disease or may induce a relapse, or it may produce a temporary arrest of the spells or even a permanent cessation. The disease is more liable to occur in the first few days of the puerperium and during lactation. Prolonged labor or forceps delivery may induce

convulsions. Convulsions occur in about one out of every five or six hundred normal pregnancies, but if acute nephritis develops they occur in about 25 per cent.

MASTURBATION or other venereal excesses may provoke a latent tendency to epilepsy. It is not infrequently associated with epilepsy, but whether it should be regarded as the cause, or whether it is another evidence of feeble cerebral inhibitory control is a matter of opinion. There is no doubt that it lowers the vitality of the brain and makes epilepsy worse.

MALNUTRITION as shown by rickets, or a general emaciation, underweight or under size, is not uncommon in epileptic children.

INFECTIOUS DISEASES.—Acute infectious diseases often cause convulsions, especially in children, and it may be stated generally that the younger the patients the more liable they are to convulsions from febrile causes. The convulsion in children often replaces the chill in adults.

Scarlet fever is frequently followed by epilepsy. The convulsions, however, do not usually come on until after the fever has subsided. *Scarlatinal nephritis* is the cause in 3 per cent. of cases.

Whooping-cough may produce cerebral hemorrhage and epilepsy. Spratling found 0.3 per cent. of cases due to this cause.

Typhoid fever is sometimes followed by epilepsy. In the cases seen by the author, there was always a history that made him suspect a typhoid meningitis and in several of them the condition of the patient indicated a previous meningitis. Chalié and Juilhe report epilepsy following typhoid fever. Out of 120 cases of epilepsy, 7 had typhoid fever according to Dide, without claiming that the epilepsy was due to typhoid fever.

Typhus fever, measles, influenza, diphtheria, smallpox, malaria, yellow fever, vaccinia, acute anterior poliomyelitis, have all rarely been thought responsible for epilepsy. *Pneumonia* is often preceded by convulsions, but is only rarely followed by epilepsy. *Meningitis*, on the other hand, is frequently the basis of persistent convulsions. *Insolation* sometimes causes epilepsy.

Syphilis.—Fraser and Watson from Wassermann tests give us reason to believe that about 50 per cent. of mentally defective and epileptic children are tainted with syphilis. These figures are higher than in the author's experience with children, but in cases developing after thirty years of age a much larger percentage is due to syphilis and, according to some writers, 9 out of 10 cases in adults are due to it. While convulsions due to syphilis occur most often in hereditary and tertiary cases, they sometimes occur in the secondary stage and are probably then due to meningeal involvement. Guenot reports a case in which the first convulsion occurred within a month after the appearance of the chancre (man, aged 26). Thomsen and others, out of 2,061 cases of mental deficiency in Denmark, found a positive Wassermann reaction in only 1.5 per cent. Out of 259 epileptics (aged 5 to 70 years), only 1 gave a positive reaction (0.39 per cent.). The coincidence is more fre-

quent in America than in Denmark. It is almost impossible to reconcile the Danish experience with that of other countries. In the writer's experience syphilis is one of the most common causes of epilepsy.

WORMS.—*Hookworm* is the commonest worm causing epilepsy in the South. Stiles has found a general invasion of the body tissues and it has not yet been proven whether epilepsy associated with hookworm is due to the direct invasion of the brain by the parasites, or to edema of the brain produced by the anemia caused by the parasites, or whether the convulsions are of reflex origin.

Tania solium or *saginata* are not uncommon causes of epilepsy which has usually been ascribed to reflex irritation. On the other hand Stiles found *Cysticercus cellulosæ* in the brain in 117 out of 155 cases. It is probable that the epilepsy is due directly to the brain infection.

Tenia echinococcus or hydatid worms existed in the nervous system in 122 out of 1,634 cases of the disease examined, according to Osler. These are sometimes calcified and persist in the brain long after the worms have disappeared from the intestine. Here again the epilepsy may be the direct result of cerebral irritation. Osler quotes Davies Thomas, of Australia, who collected 97 cases, including some of the *Cysticercus cellulosæ*, and found the cysts more common on the right side of the brain than on the left, and more frequent in the cerebrum. The symptoms resemble those of tumor, headache, convulsions, and gradually developing blindness.

Intestinal Parasites.—There is no doubt that convulsions are frequently caused by intestinal parasites, but just how this is brought about is uncertain. The following possibilities exist:

(a) The general ill health of the patient would, of course, render the nervous stability and equilibrium of the patient less.

(b) The reflex irritation may act as a factor in a summation of stimuli.

(c) Peiper has advanced the theory that intestinal parasites produce toxins.

(d) In *Tænia solium* there is often cysticercus formation in the brain, but whether in all cases of epilepsy associated with tapeworm is not known.

(e) Hookworm has been found to invade all of the organs of the body; whether or not it has ever been found in the brain in association with epilepsy is not known to the writer, but the probability of the epilepsy due to hookworm being due to its presence in the brain seems very great. It is often found that the convulsions do not cease after freeing the intestinal tract of the eggs and worms, and in order to cure the epilepsy drugs have to be given for absorption into the system in order to kill those in the tissues as well. It is interesting to note that while cysticercus occurs mostly from 40 to 60 years of age, the cases with hookworm are mostly in the early years of life.

Peachell reports a fatal case of *Cysticercus cellulosæ* in the brain with status epilepticus, although there was no tapeworm found in the intestines.

The history of having had worms should give rise to just as great suspicion of their being the cause of the convulsions as if they were present at the time of the examination.

Symptomatology.—Epilepsy may be classified from a symptomatic standpoint, from an etiological standpoint, or from a pathological standpoint. For the purpose of studying the symptomatology it may be divided into (1) grand mal or major fits; (2) petit mal or minor fits; (3) psychic epilepsy or mental attacks; (4) jacksonian epilepsy; (5) status epilepticus; (6) epileptic equivalents. (*See Clinical Varieties.*)

A complete and typical attack may be divided into five stages: (1) the aura; (2) loss of consciousness; (3) tonic spasm; (4) clonic spasms; (5) coma or sleep.

The premonition of an attack of epilepsy may consist of (A) *prodromata*; (B) *an aura*. The latter may be a sensory hallucination which may be (1) visual, (2) auditory, (3) olfactory, (4) gustatory, (5) cutaneous, (6) visceral; or it may be (7) motor, (8) psychic or mental, (9) vasomotor, (10) secretory.

THE PRODROMATA.—In some cases prodromal symptoms may occur which may exist for several hours or several days before the actual spell. Thus some patients are glum, cross, inclined to irritability, excitability, unsociable, or show various other evidences of variation from their normal for several days so that they or the family are able to predict a spell fairly accurately. Gowers says that "swiftness is an essential element of ordinary epilepsy, but this does not preclude the possibility of deliberation." In some cases there are vertigo, fullness in the head, depression or tremor, or even muscular twitchings for hours or days before the attacks come on. Pruritus, urticaria, erythema, spasms of sneezing, tinnitus aurium, photophobia, etc., are less common prodromata (Oppenheim), or delusions lasting one or several days may precede the attack (Walshe).

THE AURÆ.—The aura or premonition of an attack occurs a few seconds or a few minutes before the attack. Each patient usually has the same aura before each attack, but may not have any aura with some attacks. Very rarely does a patient start his attacks with different auræ. Sensory auræ are more common than motor auræ. Sometimes the aura occurs without being followed by an attack. This may occur spontaneously or the patient may ward off the attack. (*See Treatment.*) An aura occurs in one-half of the cases, but not in one-half of the attacks. When no aura occurs, the origin of the attack is usually in a latent portion of the brain (as in the frontal lobe), so that the patient becomes immediately dazed or unconscious and there is often transient conjugate deviation of the eyes or a sharp cry (W. A. Turner).

(1) *Visual auræ* may consist of seeing flashes of light or sparks which may be either white, yellow or red, or of moving balls of fire which may be colored. In some cases the hallucination takes more definite form, and objects, people or a hand of cards are seen which cannot be clearly remembered afterwards and seem like a dream. Photophobia occasionally occurs. Animals, as a black cat, landscapes, or other objects.

are seen moving nearer or further from the patient. There may be loss of sight, macropsia, micropsia. Transitory or paroxysmal amblyopia may precede the convulsion.

(2) *Auditory auræ* often consist of buzzing or ringing in the ears, or occasional hissing, whistling noises, roaring or loud reports like a pistol shot, rarely a word, sentence, or music. Vertigo.

(3) *Olfactory auræ*, parosmia, are usually disagreeable odors, in one of the writer's cases so foul, putrid and stinking that the patient asked that if it were possible to be relieved of only one feature of his illness, that the writer cure the odor and let the convulsions continue.

(4) *Gustatory auræ* are also apt to be disagreeable and do not conform to any of the four fundamental forms of taste in any of the cases seen by the writer.

(5) *Cutaneous auræ* consist of numbness, tingling, itching, burning, prickling, or other paresthesiæ; sometimes of painful sensations in the body or head. The paresthesiæ start in the hands, feet or face and travel upward.

(6) *Visceral auræ* consist most often of epigastric sensations (occur in 15 per cent. of the cases), which are indescribable. They ascend gradually or quickly up the chest and throat to the head, and when they reach the head the patient falls unconscious. It is probable that this is due to the distribution of the pneumogastric nerve. A relation certainly exists between epigastric sensations and a sensation of fear or apprehension. This relation is not confined to epileptic attacks, but various symptoms in the area of the vagus distribution may form a premonition or aura to an epileptic attack, such as vague indescribable feelings in the stomach, pain, or cramps, cardiac sensations, pain in the heart or palpitation, a feeling of oppression about the heart, sensations of suffocation or strangulation, esophagismus, associated with such mental symptoms as oppression, a sensation of impending death, fear, apprehension, and a desire to be alone. Occasionally there is nausea (ninth nerve) or even vomiting. Osler mentions the occurrence of boulimia in epilepsy often in paroxysmal attacks. The sensations may be hypogastric or precordial.

(7) *Motor auræ* usually start with twitching of a single muscle or group of muscles and gradually extend to other muscles in an orderly fashion, or they may consist merely in a tonic contraction of a muscle or group of muscles, but do not usually extend beyond a portion of a limb or one whole limb before consciousness is lost. Sometimes they consist of the head being rotated to one side or the eyeballs being turned to one side, or flickering of the eyelids. It is rare for bilateral motor irritation to precede an attack, and the author has never seen it. In some cases the patient runs forward or backward or in a circle, or may make scratching gestures, but these are very rare. Not infrequently children will run to their mothers crying, as if for protection, when a spell is coming on. Aphasia or dysarthria may precede the attacks. According to Oppenheim, crying, singing or whistling may rarely represent the aura or equivalent of an epileptic spasm. Yawning, singultus or in-

spiratory spasm may occur (Oppenheim). Instead of the usual aura the patient may turn around rapidly or run for a few seconds or minutes (epilepsia proeursoria).

(8) *Psychic aura* may consist of anxiety, fear, depression, or the recurrence of previous mental impressions—terror, dreams, or delusion a vague dreamy state or a sensation of strangeness.

(9) *Vasomotor aura* may consist of flushing or pallor, erythema, urticaria, ischemia or angioneurotic edema.

(10) *Secretory aura* consist of profuse perspiration, salivation or sometimes evidences of gastroenterorrhea.

GRAND MAL OR MAJOR ATTACKS.—Some patients have only major attacks, others only minor attacks, but frequently they have both. Of the cases of epilepsy 60 per cent. are of the grand mal type, while 32 per cent. have combined grand mal and petit mal.

The Attack.—Without any apparent provocative cause, such as an emotional disturbance, fight, fright, or sight of an accident, blood, etc., and with or without a premonition or aura, the patient loses consciousness and may or may not utter one sharp shrill cry, due to air being forced through the spasmodically closed glottis by the powerful contraction of the respiratory muscles, or sometimes make a gurgling noise. These symptoms occur after the loss of consciousness and the patient falls wherever he happens to be. The following events take place in rapid succession and are often not chronologically related by the lay observer. When the patient falls, there is first a tonic contraction of the muscles, the fingers and wrists are flexed, the thumbs adducted, the elbows flexed, the head retracted, the legs rigidly extended—rarely flexed; the muscles of one side may contract more powerfully than those on the other, so that the body may be curved to one side; the head or face may be turned to one side; the eyes may turn to the same side, but are usually rolled upward, the lids usually open, the pupils widely dilated; the pupil reflex is lost, the conjunctival reflex is absent, the eyeballs may seem to protrude, while in other cases the eyes are closed.

The face is usually red or cyanotic, bloated, the veins of the face and neck congested and full, with profuse sweating. Pallor may precede the redness of the face or persist throughout the attack.

Rarely the head, trunk and extremities are flexed. During the tonic stage the urine or feces may be passed. The tonic spasm may be so brief that it is not noticed at all by the observer, or it may last a few seconds or half a minute when the muscles begin to tremble and then develop into a clonic spasm, starting with short twitchings in the head, trunk and limbs, which become more and more violent, consisting of alternating contractions and relaxations, becoming more rapid at first, but with a tendency towards the close of the convulsions to become more powerful but slower and slower till they cease altogether. The eyeballs jerk in the direction of the strongest contractions. The convulsion may be unilateral or start as a unilateral convulsion and then become bilateral.

The attack in detail is as follows:

Unconsciousness is the most characteristic feature of epilepsy and occurs in all cases of grand mal during the whole of the attack in most cases, or at least during a part of the attack in all cases.

Color.—At the very onset of the attack there is pallor, but with the rigidity of the chest walls and cessation of respiration this gradually changes to cyanosis and as the attack progresses congestion of the veins of the face, neck and head occurs and the complexion becomes a dusky purplish red, although in some attacks the patient is pale during the whole attack.

The Cry.—The cry occurs in only a small percentage of the attacks, and occurs after the loss of consciousness. It consists of one sharp, shrill cry, and is never repeated during the course of the attack, and the patient has no memory of it afterwards.

The Tonic Spasm.—Usually no history is obtainable from the family in regard to a tonic stage and their description usually starts with the fall and is followed by a description of a clonic stage only. Often the tonic stage lasts only a second or two, though in other cases it may last longer, but it is always short in proportion to the clonic stage. In the tonic stage it may consist merely in the drawing of a single muscle or a group of muscles, the flexion of a finger or the toes, or may march up a whole extremity or affect a whole half of the body either by marching in an orderly fashion, or may involve the whole body at the same time. The march follows the anatomical location of the centers in the brain, and if it starts in the face it extends first to the arm and then the leg, and if it extends to the opposite side of the body it will then affect the leg, then the arm and then the face. In the majority of attacks both sides are affected apparently at the same time or at least in such rapid succession that no order of march is observed. In the cases which start in a certain part of the body and show an orderly sequence of tonic spasm, this same characteristic is preserved in each succeeding attack. Usually in the tonic stage the wrists are flexed, the thumb is adducted and lies in the palm with the fingers grasping it, or the fingers may be extended, the elbows flexed, the arms raised, the back arched (opisthotonos), the head drawn backward, or if the convulsion is stronger on one side than the other, the head, eyes and mouth are drawn toward that side. The legs are usually rigidly extended.

While the above description of the position of the body and extremities is the usual one, any position may be assumed, and the writer has seen cases in which the body was flexed, the head forward, the chin resting on the chest, one arm and one leg flexed, while the other arm was drawn across the chest and the other leg extended, and in the same case in other attacks other positions were assumed.

The Eyes.—There is usually a conjugate deviation of the eyes in the direction the face is turned, or the eyeballs are rolled upward so that only the white shows; the lid fissures are open, the pupils are widely dilated and do not react to light, and the conjunctival reflex is lost. According to Barker, the pupils in this stage are contracted, while

Gowers says that although initial contraction at the beginning of the tonic stage had been reported he had never seen any cases that were not dilated, and this has been the writer's experience also. Certainly after the attack is well started the pupils are always dilated and Spratling found that this dilatation took place even before any muscular movements occurred. The eyelids are usually open, though sometimes closed. The dilatation of the pupils is probably due to the sympathetic nerve. That it is not due to asphyxiation is shown by its occurrence at the beginning of the attack and in petit mal. Stewart says the mydriasis ceases at the completion of the tonic stage, then the pupils become contracted and remain so until the stage of coma has passed off—or sometimes hippus may occur.

The urine may be passed in the attacks. Many cases never pass urine during the attacks, while others give a history of passing it in some of their attacks, but only a minority of them. It is apparently more frequent in children than adults, and bed wetting (nocturnal enuresis) is always suspicious of epilepsy.

Defecation during the attacks is much less frequent than urination, and when it occurs is usually associated with urination at the same time. As in the case of urination, the percentage of attacks in which it occurs is much smaller than the percentage of the number of patients in which there is no history of it.

Sweating is often profuse and general, so that the patient is wet all over. It is sometimes greater on one side of the body than the other.

The Clonic Stage.—After the tonic stage has lasted a second or more, or even without being obvious, the clonic convulsion starts with alternating contractions and relaxations of the muscle; the convulsion usually starts in the same part of the body in which the tonic spasm started and follows the same line of march, or in cases in which the tonic spasm was general and bilateral the clonic convulsion starts in the same way. The jerking of the muscles is usually slight at first but becomes stronger and faster till the end of the attack, when it becomes slower and more violent until it ceases altogether. During the clonic stage, the alternating contractions and relaxations of the respiratory muscles lead to a lessening of the cyanosis, the respiration is rapid and noisy and may be accompanied by groaning or moaning noises; there are both increased flow of saliva and failure to swallow or expectorate it, so that it is worked into a froth ("foaming at the mouth") by the forced passage of air through it. The tongue is sometimes bitten in the tonic stage, but more frequently it is bitten in the clonic stage, in which it may be chewed repeatedly on the side towards which it deviates, this being the same side towards which the face and eyes are turned and on which side the general convulsion is stronger. The biting is due to the jaw muscles being alternately contracted and relaxed and the tongue being drawn to one side. The biting of the tongue may give rise to fluid which makes the foaming at the mouth bloody. There is usually profuse general perspiration, often urination, rarely defecation, and still more rarely ejaculation of seminal fluid. The clonic stage lasts

from a minute to ten minutes—rarely longer—and is followed by sleep or coma which may last from a few minutes to a few hours. Occasionally the patient may open his eyes after the convulsion, before going to sleep, but does not seem to be conscious. The sleep is apparently due to exhaustion from the attack and is apt to be longer after more severe attacks, and shorter after very light ones. When aroused from this sleep (which is sometimes impossible), the patient may get up and do various apparently purposive acts, or be violent with no knowledge of it afterwards—or if really awake is apt to be cross, irritable and suffer from headache. Vomiting may occur during the attack or after it, and may lead to strangulation of the patient. Polyuria or nycturia may follow the attack.

Postepileptic State.—The attacks are followed by more or less general exhaustion of the cortical motor centers, and may be more marked on one side than the other, giving rise to monoparesis or hemiparesis, which is most marked and more prolonged in cases where a focal brain lesion exists, as in jacksonian epilepsy. Sometimes the attacks are followed by aphasia or stuttering or transitory paraplegia; amaurosis and amnesic aphasia have been observed. Patients are usually rather dull and stupid after spells, but occasionally as soon as the convulsion is over, without sleep or coma, the patient gets up and seems to feel as well as usual.

Oppenheim mentions concentric narrowing of the field of vision and disturbances of general sensibility after the attacks, also erythematous flushing, tremor, conjugate deviation of the eyes, paralysis of the ocular muscles, blindness, deafness, vomiting, diarrhea, and other gastric disturbances, polyuria, salivation, edema, cutaneous emphysema, etc., as postepileptic symptoms.

PETIT MAL.—Minor epilepsy or petit mal may consist merely in loss of consciousness with or without falling and with or without motor twitching, or may consist merely in an aura such as occurs in major epilepsy without any convulsion developing. The attacks are not usually preceded by an aura. They come on suddenly and are of short duration, from a second to half a minute. They may or may not know of the attacks, which may consist in a sudden suspension of conversation which is taken up where it is left off when the attack has passed off, or sudden lapses in action which are quickly resumed again. During the attacks there are a change of facial expression, widening of the palpebral clefts, dilatation of the pupils, pallor or more rarely flushing of the face, slight twitching of the muscles of the face, the lips or eyelids, tasting or smacking movements; sometimes the face is turned to one side as if looking over the shoulder, or the patient jabbars unintelligibly; occasionally there is a slight twitching of one or more of the extremities. There is no real convulsion, no stertor, no foaming at the mouth, though the patient may spit. The tongue is not bitten. Occasionally the urine is passed involuntarily. In the attack the patient may drop things from his hands or the legs may give way and the patient may fall.

Buzzard records cases of "embryonic epilepsy" which may be recognized by the sudden onset when the patient is feeling well, and in each

case each attack is identical with the others, the symptoms differing in different cases, very slight in character, and if untreated lead eventually to a distinct epilepsy. He regards the slight jerks just before dropping off to sleep, or momentary twitchings, as being frequently symptoms of the embryonic stage of epilepsy.

Sometimes the attacks are represented by night terrors, by screaming, talking in sleep, and the patient cannot be awakened until the attack is over. Somnambulism must always be looked upon as suspicious of epilepsy.

Under epileptoid signs Barker mentions (1) petit mal, (2) epileptic vertigo, (3) epileptic nightmare (*pavor nocturnus*), (4) epileptic bed-wetting (*enuresis*), (5) epileptic anxiety attacks. He says:

"In epileptic vertigo there are apparently causeless sudden attacks of transitory vertiginous feelings often combined with pallor but without loss of consciousness or falling, sometimes associated with tachycardia or bradycardia, flushing of the face, dilatation of the pupils, sweating and headache." (Barker.)

Tinnitus aurium may occur and the patient only realizes that the noise has ceased but does not realize it during its presence.

PSYCHIC EPILEPSY.—Psychical epilepsy, which is also spoken of as "para-epilepsy" (Dana) or "epileptic equivalent," or "epileptic automatism," gives rise to attacks in which, with or without prodromata or an aura, and without any apparent cause, the patient shows mental excitement, violence, fury, may commit crimes without provocation or choice of the person against whom the crime is committed, or destructive, purposeless acts, or perform various apparently purposive acts which may seem to the observer either senseless or sensible. These attacks terminate spontaneously in a second or may last for hours or days, and on recovery the patient knows nothing of what has happened during the attack.

One of the writer's patients walked forty miles to a neighboring town and, on recovery from the attack, knew of only two facts which he could associate with the trip, namely: on entering the hotel here to keep an appointment, consciousness had ceased at the door of the hotel, and it had returned while he was standing on the doorstep of a house in the neighboring town, his hat in his hand, while a lady instructed him how to reach some point in the town, although he knew nothing of what he had asked her. He had suffered from frequent epileptic convulsions and this one was replaced by psychical epilepsy.

Price reports a case in which the patient was in a confused hallucinatory and delusional state and always believed a strange man was in the room with delusions of marital infidelity. The attacks lasted 15 to 60 minutes and were followed by headache and somnolence, with no memory of the attacks.

Psychic epilepsy may consist of drowsiness, stupor, dreamy states or feelings of unreality, apprehension, dread, fear, nervous sensations, numbness, coldness, nausea, cephalic sensations, which are frequently accompanied by sensations of a vague, indescribable character. The patient may not lose consciousness, but a dreamy state may exist with a feeling of unreality (Turner).

Psychic epilepsy is rarer than grand mal, petit mal or jacksonian epilepsy.

The psychic complications of epilepsy are unconsciousness or alterations of consciousness, epileptic twilight states, progressive mental deterioration, psychoses.

Epileptic twilight states may precede, replace or follow the attacks, and may last for minutes, hours, days, weeks or even months (Barker), and in them the patient may take long journeys (ambulatory automatism, poriomania, epileptic fugues). The epileptic acts in the twilight state are often violent (murder, incendiarism, rape, etc.).

In a case of the author, just as he was about to make an ophthalmoscopic examination, the pupils suddenly dilated, the face became expressionless, the palpebral fissures widened, while the patient suddenly unbuttoned his vest and trousers, put his hand in his pocket and withdrew his knife, opened the large blade and grasped it as if with the intention of stabbing. His muscles then gradually relaxed, his shoulders drooped, he folded his knife and put it back in his pocket, his expression became natural, and he had no knowledge of having had the attack when questioned about it.

Briand and Salomon have reported cases of epilepsy with erotic impulses as fetichism for handkerchiefs and silk in a man and woman respectively.

Postepileptic sleep may be replaced by defects of memory and confusion, or pronounced mental disturbances.

In psychic epilepsy there is a suspension of mental control in which the patient is in a dream-like state resembling dual personality and of which the patient has no knowledge after the attack is over. The epileptic equivalents often occur as a substitution for motor epileptic spells, but are more common with petit mal than grand mal attacks. The same statement is true for postepileptic automatism. Mental disorders often occur when a patient is aroused after a fit and not allowed to sleep, and may follow attacks of petit mal.

JACKSONIAN EPILEPSY (*Secondary Epilepsy, Cortical Epilepsy or Focal Epilepsy*).—The contractions begin in a definite muscle or group of muscles and extend gradually to other muscles, and eventually the whole body may be involved. They may or may not be accompanied by a loss of consciousness. The convulsion may be confined to one extremity or one-half of the body. The order of involvement of the different muscle-groups depends upon the situation of the lesion and the an-

atomical relation of the centers in the brain. For example, an attack starting in the leg passes to the arm, then to the face; if it travels to the opposite hemisphere, it would involve the leg, then the arm, then the face of the opposite side. There is localized muscular weakness after the attack. The attacks may be tonic or clonic or both.

Jacksonian epilepsy is usually a sign of circumscribed irritation in the brain (trauma, tumor, abscess, gumma, tubercle, local meningitis, localized edema of the brain, vascular lesion, scar), but similar convulsions are occasionally observed in diffuse irritations, as increased intracranial pressure, in toxic states and in idiopathic epilepsy. Jacksonian epilepsy may be combined with grand mal or petit mal. Rulf cites four cases in one family of familial cortical epilepsy (jacksonian) without loss of consciousness.

STATUS EPILEPTICUS.—Status epilepticus is known to the French as "*état de mal*," and is sometimes spoken of as "acute epilepsy." In it one spell follows another in rapid succession, without the regaining of consciousness. When it occurs, it usually follows grand mal attacks and is especially liable to occur in cases which present the type of "serial epilepsy." It sometimes follows petit mal or psychic epilepsy, but in the latter cases the muscular twitching may be very slight while the coma is profound, with elevation of temperature, and the patient may die in a few hours.

According to Clark (*see* Spratling, Bibliography) there is no limit to the number of status periods that may occur before death supervenes. As a rule, three or four status periods in idiopathic epilepsy cause death, while innumerable periods of status epilepticus unilateralis or partial epilepsy may occur in hemiplegic epileptics and death not supervene.

Probably a third of the fatal cases of status epilepticus are due to the sudden withdrawal of sedatives. Smith reports a fatal case of status epilepticus in a girl 8 years old who had 1,649 grand mal attacks in four and a half days, 573 being the maximum for 24 hours. Clark says that Leroy reported 488 attacks in 24 hours and 1,000 in three days in a case of status epilepticus with fatal result. As a general rule, the convulsive stage lasts from 8 to 10 hours to 3 or 4 days—and this is followed by a stage of exhaustion and coma which may be of still longer duration. "The duration of status in its entirety is usually not less than 20 hours or over 12 days, if both stages are present." (Clark.)

About a third of the fatal cases have no real stage of coma, as they die during the convulsive stage.

MENTAL CHARACTERISTICS.—Clark found the "make up" (mental characteristic is no doubt meant) of *hemiplegic epilepsy* identical with that of essential epilepsy. The patients had "high volatile explosive temper," were egotistic, self-centered and sensitive. The paternal parent was usually quick-tempered, the mother usually mild and docile. In hemiplegic epileptics the mental deterioration consists of slowness, impaired memory, dissatisfaction and lack of interest. They

are backward at school, slow to grasp facts. Emotionally they are quick and impulsive. The power of concentration and observation is fair. They have good common sense and self-reliance. They lack imagination and originality in play. They show only a slight tendency to bashfulness and shyness. They are usually neat, clean and orderly. They are domineering and are not popular with other children; stubborn and difficult to handle; usually trustful and not suspicious; sensitive, easily offended and see slights when none are intended. They are irritable especially just before attacks. Some show anxiety and forebodings. They are usually truthful and are overserious.

Epileptics are usually irritable, selfish, self-willed, impulsive, egotistical, unreasonable at times and hard to get along with. Ninety per cent. of them show impairment of recent memory.

"The epileptic character results from a progressive change in the personality." (Barker.) There are irritability, outspoken ethical defects, violent egotism, and lying. There is a tendency to violence and they often fight with slight provocation and may perform criminal acts. The intellect eventually suffers if the spells are frequent, and there are failure of the memory, enfeeblement of judgment, mental slowness, stupidity, and finally dementia. Epileptics show a marked intolerance for alcohol.

There is no memory of the attack (amnesia) and many patients have attacks without knowing it, while others have learned from the way they feel after the attacks to surmise the fact that an attack has taken place.

Healy found (a) a strange variability in mental capacities and changeable moods and dispositions; (b) the epileptic character, emotionalism with marked inconsistency shown in their feelings, irritability, sudden anger and vicious conduct without apparent cause, sullenness, self-love, defective appreciation of the rights of others, obstinacy; (c) mental deterioration, perception and will are affected, finer ethical discriminations are lost, and moral inhibitory powers are lowered.

INTELLIGENCE.—Many very brilliant men have had epilepsy—Mahomet, Swedenborg, Julius Cæsar, Peter the Great, Napoleon, and many other leaders of men have shown some special ability, which in itself has sometimes been classed as evidence of abnormal brain development, namely, as approaching the class of idiot savants. Often the struggle, determination, and accomplishment of the desire to excel is a defense reaction on the part of men who, realizing their deficiencies, fight all the harder for supremacy, while others of a normal, balanced mind find more ease and comfort in a middle path in life.

In considering the intelligence of epileptics two facts must be kept clearly in mind: (a) that mentally defective children are far more frequently afflicted with epilepsy than are normal children; (b) that frequently repeated epileptic attacks usually lead to mental impairment. Most epileptics show some mental peculiarities. They are especially irritable, hard to live with, want their own way about everything, are often suspicious and excitable, and extreme restlessness is often noted

in children. Frequently repeated attacks over long periods of time lead eventually to enfeeblement of the intellect, failure in memory and eventually dementia. Personality often determines the direction of mental deviation, so that we may have hypochondria or actual melancholia in some patients, while acute mania develops in others. Mental diseases follow either major or minor spells.

PHYSICAL FINDINGS.—Epileptic Facies.—In the majority of cases there is nothing in the physiognomy to suggest epilepsy, but sometimes, and especially in old cases, there is something about the facial expression that leads us to ask if the patient has ever had convulsions. The most easily described picture is the appearance of mental deficiency—a heavy look about the face, a vacant expression about the eyes, a rather deep far-away look, often with large pupils and wide palpebral clefts, often thick lips, and if these are associated with acne on the face (which suggests the use of bromids) and scars on the face (which suggests falls), we may suspect the disease, but there is never anything about the facies that forms conclusive proof of the disease.

Scars.—The tongue, on the other hand, will sometimes give us much safer ground for a diagnosis, from scars produced by biting the tongue, but this never precludes the possibility of the tongue having been bitten from other causes. Scars on the tongue occur in 22.5 per cent. of cases.

The presence of burns occurs in 12 per cent. of cases. Dislocations, especially of the shoulder-joint or jaw, may be present, or contusions, petechial subcutaneous hemorrhage, or conjunctival hemorrhage.

Temperature.—According to Barker, during the attack the temperature rises slightly, not more than 0.5° C. (0.9° F.).

Spratling found slight fever immediately after attacks of grand mal in 57.5 per cent. of cases and subnormal temperature in 15.5 per cent. Half an hour after attacks, 66 per cent. showed fever (highest 102.2° F. [39° C.]), and an hour after the fit 64 per cent. showed a rise of temperature from 0.2° to 3.5° F. ($.36^{\circ}$ to 6.3° C.). Even after petit mal attacks he found fever in 60 per cent. (immediately) and 70 per cent. one-half hour after the fit and in psychic attacks the results were almost identical. In status epilepticus he found the temperature varied from 102.4° to 107.5° F. (39.1° to 41.9° C.). In serial epilepsy the highest temperature was 105.8° F. (41° C.), caused by 140 convulsions, while in the other cases none went above 100.8° F. (38.2° C.).

Pulse.—Russell found the pulse in a fit feeble or absent. The writer has found the pulse feeble in some attacks when the heart on auscultation was pounding very forceably, and was very rapid. On the other hand, epilepsy is said by Osler to be associated at times with a very slow pulse. "Cardiovascular epilepsy is usually a manifestation of advanced arteriosclerosis, and is associated with a slow pulse." (Osler.)

Several writers (Russell, Hare and others) have been so fortunate as to have their fingers on the pulse at the very moment of onset of the attacks and observed that there was a cessation of the pulse, while Hebb was listening to the heart (in aortic regurgitation) when it suddenly stopped beating, being followed by a general convulsion.

Gibson, Good and Penny obtained tracings of the pulsations of the brachial artery immediately preceding the fit in five epileptic attacks, using the Erlanger's sphygmomanometer. The tracings show that up to the time when the movements of the arm affect the tracing there was no cessation of the pulse, and in all instances there was a gradual increase in the rate of the pulse, culminating in the onset of the fit.

Respiration stops in the tonic stage and becomes very rapid and noisy during the clonic stage. Between attacks there is no alteration in the respiration.

Weakness.—Muscular weakness may exist in the form of monoplegia, hemiplegia, or even paraplegia. This is most marked and most persistent in cases with organic brain lesions, but in any case the attack, if more marked on one side, may be followed by weakness of the muscles on the side of the strongest convulsion, and is associated with a diminution or loss of cutaneous sensibility, which is temporary. Temporary amblyopia, deafness, impairment of smell and taste may also occur.

According to Redlich, the left-handedness common to epileptics may result from a previous brain disease (quoted by Oppenheim).

The weakness after epileptic attacks is only transient, while a persistent localized weakness indicates organic brain disease. More or less general exhaustion follows all severe attacks.

Reflexes.—Audenino finds that the so-called essential antagonism between cutaneous and tendon reflexes is present in 66 per cent. of his cases during remission. In 51 per cent. the tendon reflexes were increased and one or both cutaneous reflexes diminished or absent with the reverse condition in the remaining 14 per cent. Asymmetry of either cutaneous or tendon reflexes was present in 73 per cent. of his cases. Before attacks the cutaneous reflexes are increased, the tendon reflexes diminished. In the postepileptic period the abdominal cutaneous reflexes are absent, the tendon reflexes increased, and the plantar responses are either extensor or absent.

According to W. A. Turner, after a major fit the knee-reflexes are first abolished, later exaggerated, the abdominal reflexes are lost, and the plantars show a temporary extensor response. These signs are more marked after severer convulsions.

Barker finds that the tendon reflexes are sometimes abolished during the attacks and the Babinski sign may be positive.

In epilepsy the Babinski sign disappears shortly after the attack.

According to Oppenheim the reflexes are not obtained during the attacks and the tendon reflexes are absent for a short time after the attacks. Between attacks they may be normal or exaggerated. The Babinski toe-reflex is said to be present during and a short time after the attack. The "feed-reflex" is sometimes present.

In several cases the author has found the knee-reflex exaggerated during the convulsion.

Lesieur and others found the oculocardiac reflex always considerably exaggerated in epilepsy, with marked slowing of the heart (12 to 50 beats per minute).

Graetz regards spasmophilia as a constitutional defect characterized by (1) increased excitability to anodal stimulation; (2) the occurrence of Chvostek's sign; (3) direct muscular irritability (with tumor formation on percussion); (4) hypertonus of the arteries from overaction of the vasoconstrictors; (5) alteration in the blood state (presence of Pappenheim's leukoblasts, nucleus tending to polymorphism, with basophile cytoplasm) and increase in large mononuclears. He finds that most epileptics show these signs of spasmophilia.

X-ray Examination.—X-ray examination shows in some cases kinks in the colon, malformations, adhesions, extensive dilatations, colonic stasis, adhesions of the appendix with ileocecal valve insufficiency, etc., but whether these occur more often in epilepsy than in other diseases has never been worked out.

Johnson in studying the skull of idiopathic epileptics found hyperostosis involving the clinoidal processes, the base of the anterior fossa, and in some instances the postclinoidal region. In some cases the anterior and posterior clinoidal processes meet.

M'Kennan, Johnston and Henninger in 95 cases found that 7 were due to pituitary tumors. In these tumor cases there was local thinning of the body of the sphenoid or the clinoidal processes from pressure, instead of local thickening or overgrowth.

LABORATORY FINDINGS.—*Urine.*—Intestinal putrefaction is closely related to the attacks. There is an increase in indican just before and during the epileptic fit in many cases.

Injection into animals of urine taken after the attacks shows it to be more toxic than normal urine; between attacks it is hypotoxic.

Hallager says albumin is always present for a short time in the urine after an attack, which he attributes to a temporary anemia of the kidney due to vascular constriction, and this is followed by a reactive dilatation of the vessels with polyuria.

Gowers, on the other hand, found albuminuria extremely rare, but found an increase after fits in a case of nephritis.

Garnett has used the complement-fixation test in epilepsy. The urine of the epileptic was used as the antigen, and the serum of the epileptic as the antibody. Normal urines and sera were used as controls. He concluded that the serum of epileptics contains some substance of the nature of a specific antibody, and the urine of epileptics generally contains a substance specific to the antibody in the serum. The serum of non-epileptics does not contain this antibody. The urine of non-epileptics occasionally contains a substance which, when mixed with the serum of an epileptic, is capable of deviating a small amount of complement, but not to the same extent as when the urine of an epileptic is used.

The urine after an attack contains an increased amount of urea, nitrogenous elements and phosphates.

Voison and Peron found a hypotoxic condition of the urine before and a hypertoxic condition after the fit.

Blood.—Clark finds the blood toxic after attacks, as shown by the effect of injections into animals.

Shaw states that there is a diminution in the *alkalinity* of the blood.

Donath found *cholin* in the blood and cerebrospinal fluid, and it has also been found in general paralysis of the insane with epileptiform attacks. Others regard it as a result of the attacks. The injection of large doses of *cholin* into the circulation causes convulsions and paralysis.

John Turner states that the average rate of *coagulation* of the blood is quicker in severe cases of epilepsy, especially in serial epilepsy, while the patient is having fits; that there is a further quickening of the coagulation rate up to 24 hours before a fit; and that from 24 to 48 hours after a fit there is a retardation in the rate of coagulation. Besta found that in 37 out of 45 cases of epilepsy the coagulability of the blood is diminished. Intravenous injections of calcium chlorid cause contraction of the pupils and a reduction in the irritability of the papillomotor fibers of the cervical sympathetic nerves, which is in contrast to the condition existing in epilepsy.

Brown found the average *viscosity* of the blood in 15 cases of epileptic insanity to be 4.8 times water, as compared with 6 healthy persons in whom the viscosity was 4.2 times water. In 46.6 per cent. of the cases of epilepsy the viscosity was above 5, while one of the 41 other cases examined showed a viscosity higher than 5 times that of water. The viscosity in epilepsy rises just before a fit and is highest in cases having frequent and severe convulsions.

According to Elsner, Fockenheim stated that before an attack there was "a marked reduction of *hemoglobin*, the destruction of both red and white cells, and these conditions with increased coagulability of the blood he believes are of diagnostic and prognostic importance. . . . The large mononuclear leukocytes are increased before the attack. There is *poikilocytosis* and *microcytosis* during the attack." Spangler in 100 cases found the average hemoglobin 84 per cent.—the red and white cells about normal between attacks; but there is uniformly a tendency to leukocytosis or hyperleukocytosis at the time of, and frequently for twenty-four hours after, the attacks. The differential count was normal, except that the large mononuclears averaged 9 per cent. above the normal range. The coagulation time is shortened before an attack and was shorter between attacks than in normal people. The alkalinity of the blood was found lower in epilepsy than in control individuals on the same diet.

Ceni has introduced a theory of autocytotoxins and anti-autocytotoxins in the blood of epileptics, but his theory has not been confirmed.

Gordon found that one drop of blood taken from an epileptic and dropped into fifteen drops of cerebrospinal fluid of another epileptic resulted in complete hemolysis, and vice versa, while a drop of blood from a patient added to his own cerebrospinal fluid remained coagulated for days.

Cerebrospinal Fluid.—The occurrence of *cholin* in the cerebrospinal fluid after epileptic attacks is reported by Donath.

Thabius and Barbe found the chlorids increased and glucose diminished.

An overgrowth of the anterior and posterior clinoidal processes is often found in cases of petit mal developing between the ages of fifteen and twenty-five years, and accompanying this gradual growth there is a steady increase in the frequency and severity of the attacks. Hypopituitarism is the supposed cause of the epilepsy in these cases.

Diagnosis.—*Convulsions* occur (according to Leftwich, "Index of Symptoms," 1915) in the following diseases:

Abscess of brain	Hysteria
Absinthism	Idiocy
Acute yellow atrophy	Indigestion
Addison's disease	Infantile hemiplegia (onset)
Ague (cold stage in children)	Infantile paralysis (onset)
Alcoholism (chronic)	Intussusception
Anemia of brain	Irritating scar
Aortic stenosis	Jacksonian epilepsy
Apoplexy (cortical)	Jaundice
Apoplexy (unilateral)	Lead poisoning, chronic
Asphyxia (term)	Malingering
Cerebrospinal meningitis	Meningitis
Cirrhosis of kidney	Myelitis, acute
Coal-gas poisoning	Myoclonus epilepticus
Compression of brain	Nephritis, acute, third stage
Cysticerci of brain	Pachymeningitis, cerebral
Dentition	Pneumonia, acute, in children
Disseminated sclerosis, third stage	Poisoning by arsenic, alcohol,
often unilateral	brucin, hydrocyanic acid, picro-
Encephalitis	toxin, strychnin, tobacco, nar-
Enteritis, acute, in children	cotic irritants in general. In
Epilepsy	children: atropin, morphin or
Ergotism	santonin.
Exanthemata (onset)	Pontine hemorrhage
Exostosis of skull	Pregnancy
Frights	Puerperal state
General paralysis of insane	Rickets
Heart disease, congenital	Round-worms and tapeworms
Hematoma, dural	Salvarsan poisoning
recurrent	Softening of brain
Hemorrhage	Spina bifida, when about to burst
Hydrocephalus, chronic	Spinal meningeal hemorrhage
Hydrocephalus, spurious	Starvation
Hydronephrosis	Status epilepticus
Hydrophobia-tetanol	Status lymphaticus
Hyperemia of brain	Stokes-Adams' disease
Hyperpyrexia	Sunstroke
Hypertrophy of brain	Syphilis, cerebral

Syphilis, hereditary
 Syphilitic nodes
 Tetanus
 Thickening of skull

Thrombosis of brain
 Tumor of brain
 Uremia
 Ventricular hemorrhage

The diagnosis of epilepsy rests upon the *age of onset* and a *history of the attacks* in the majority of cases; therefore, an accurate description given by some one who has seen the patient have an attack is necessary. The patient's description is of little value, as he is unconscious during the attack, but the evidence he may give may in some cases be quite sufficient, such as having found himself lying on the floor with saliva on the cheek, the tongue bitten, the urine passed, general muscular soreness, etc. One attack is never sufficient to conclude that the patient has epilepsy, as it may be only a symptomatic convulsion from some other disease. Indeed, convulsions are not a disease, but merely a symptom of many different diseases (about 100); therefore it is only when they form the chief and most characteristic feature of the complaint, occur repeatedly, without obvious provoking cause, the patient being well between attacks, that they should be grouped as "epilepsy." Or, on the other hand, when repeated spells of unconsciousness with the well-known symptoms (*petit mal*), or repeated sensory disturbances are described by the patient (aura or sensory epilepsy) or repeated psychic disturbances (psychic epilepsy) occur, the patient being in good health or his usual normal state between attacks, we feel justified in making a diagnosis of epilepsy, and then only when we have excluded all other diseases in which convulsions occur and in which, instead of the disease belonging to the attack, the attack belongs to the disease (as brain tumor, general paralysis of the insane, multiple sclerosis, etc.). A thorough knowledge of epilepsy should first be acquired and then a consideration of the leading features of other diseases in which convulsions occur should be considered. There is usually no difficulty in diagnosis if there is a history of previous attacks, a good description of the attack, or if the physician sees the case during the attack. The history of usual good health, the sudden onset without apparent cause, with or without an aura, the loss of consciousness, the tonic spasm, the jerking of the muscles, the open eyelids, the position of the eyeballs, foaming at the mouth, biting the tongue, the color of the face, the passage of urine or stools, followed by coma and sleep and a normal condition following the attack, are quite characteristic. There are cases, however, which are not typical, and where no history is available, no one present who has seen the attack, and these may present difficulties.

In *petit mal* and psychic epilepsy the diagnosis must rest chiefly upon the sudden, brief loss or impairment of consciousness, occurring repeatedly. Oppenheim says the "absence of the reflexes, even of the tendon-jerks, is a proof of the genuineness of the attack."

DIFFERENTIAL DIAGNOSIS.—The disease must be differentiated from:

(1) *Infantile Eclampsia*.—Most of the cases of convulsions in childhood are not to be regarded as true epilepsy. Some writers prefer to

use the term *infantile eclampsia* for cases in which convulsions usher in acute infectious diseases, cases due to gastric and intestinal disturbances, worms, adherent prepuce, dentition, rickets, physical or psychic trauma, etc. Only one out of eight of the cases of convulsions in infancy persist as epilepsy. It is rather immaterial whether we choose to make a distinction between infantile eclampsia and epilepsy or not. They both indicate a defective development of the inhibitory apparatus, or nervous instability, and it is more a matter of prognosis than diagnosis as to whether the child will "outgrow" the convulsions.

The tendency to spasmophilia is certainly greater in children than in adults and tends to become less with increasing years, but it is more marked in the feeble-minded than normal individuals. In infantile eclampsia the attacks are apt to assume the form of serial attacks, and in them the tonic and clonic stages may be mingled, going from one to the other without the orderly sequence of true epilepsy. There is no clear distinction between infantile convulsions and epilepsy.

(2) *Apoplexy*.—According to Osler, "in more than 50 per cent. of the cases of infantile hemiplegia the affection follows severe convulsions."

The differentiation of apoplexy from epilepsy will rest upon the history of the case, the presence of plausible causes for apoplexy, the unilateral convulsion (which may, however, be general, or also unilateral epilepsy may occur), the evidences of paralysis (which are transient in epilepsy, more permanent in apoplexy) and the failure to return to normal as occurs in epilepsy. The usual physical signs of apoplexy should be looked for. Epileptic convulsions are much less common after apoplexy in adults than after apoplexy in children.

(3) *Hysteria* offers probably more difficulty than any other disease, although in the vast majority of cases there is no difficulty at all. The chief source of error is insufficient or defective data upon which to base an opinion. According to W. A. Turner, "both epilepsy and hysteria are diseases founded upon a hereditary degenerative endowment." According to Janet the "fundamental characteristic of hysteria is a dissociation or severance of some mental processes from the main personal consciousness." The chief points are that hysterical spells are most apt to occur before or soon after retiring at night and are usually preceded by an emotional cause, such as a quarrel, disappointment, fright, shock, etc. The premonition often consists of palpitation of the heart, depression, malaise, choking sensations, dyspnea, etc., often lasting for some time before the attack develops. There is often crying or laughing during the attacks and even talking from time to time, none of which occurs in major epilepsy, except one sharp cry at the beginning of some epileptic attacks. Consciousness in hysteria is not completely lost and the attention of the patient can be attracted by a stranger or by spectacular means. The eyes are not usually fixed and look about at people from time to time, the pupils are not dilated and the pupillary and conjunctival reflexes are not lost. In the hysterical convulsion there is no brief tonic spasm, followed by a clonic convulsion of short duration, but a struggling, throwing about of the arms and legs, rigidity,

opisthotonus, occasional retraction of the head, periods of rest, then more convulsions, biting of the lips, hands, bed clothing or other people, but not the tongue. Urination and defecation do not occur in hysteria. The attack usually lasts longer than epileptic attacks and can be terminated by smelling ammonia, injection of apomorphin, a few sharp words, slapping, pouring cold water on the patient, etc. In one of the writer's cases the patient jumped out of the window while the writer was heating an iron to singe the bottom of the feet. Hysterical spells usually occur at home; in the street only on patients witnessing accidents, etc. It is extremely rare that a patient is ever injured in a hysterical convulsion. Between attacks, the mental dullness of epileptics is rather in contrast to the brightness and emotional nature of hysterical subjects. Hysterical patients usually find a convenient and comfortable place for the attacks. There is no Babinski toe-reflex during or after hysterical attacks. The persistence of the Babinski phenomenon for several hours after the attack is indicative of symptomatic epilepsy. Painful pressure upon the supra-orbital nerve or upon the ovaries will produce evidences of consciousness in hysterical subjects. Even with these criteria before him, there are occasional cases in which a physician finds himself unable to make a definite diagnosis. There have been cases recorded in which the pupillary reflex was not lost in epilepsy, and in which it was lost in hysteria, but a hysterical subject would never have all of the evidences of epilepsy and none of the evidences of hysteria, so that a diagnosis, when the attack is witnessed by the physician, could hardly leave any question of doubt. It must, however, be said that both diseases may exist in the same patient. In hysteria there is no tendency toward dementia. Important also for the demonstration of hysteria are the anatomical and psychic bases upon which hysteria is produced, namely, lacerations of the perineum and cervix, cystic ovaries, displacement of abdominal and pelvic organs, a sexual excitement without gratification, sexual abuses, social environment, drunkenness or irregular late hours of the husband, unkindness in the home, fears of punishment or exposure, etc.

(4) *Alcoholism*.—Convulsions occurring in alcoholism are distinguished by the history, age of onset and evidences of alcoholism.

(5) *Syncope*.—Syncope usually has an apparent cause, occurs from exhaustion, sudden changes of position, heart disease, the sight of blood or an accident, etc. There are pallor, sweating, feeble pulse, feeble heart action and no convulsion. It could be confused only with the rare epileptic attacks which consist merely in a fall and unconsciousness without convulsions, but all repeated and causeless attacks of syncope should create an investigation and thought of epilepsy. Syncope is not periodical. There are retention of the pupil and other reflexes.

(6) *Meningitis* is distinguished by its usual symptoms and signs and should not give rise to any confusion.

(7) *Brain tumor* may develop in epileptics, or convulsions may be merely one of the symptoms of brain tumor. The usual symptoms and signs of brain tumor should invariably be looked for in cases of con-

vulsions developing after thirty years of age. Convulsions may occur from a growth anywhere in the brain and may be general in character, though more often jacksonian. They occur as early symptoms of brain tumor especially in children. Turner says that convulsions may precede the symptoms and signs of intracranial tumor by many years. In favor of tumor are a well-defined local warning, some degree of post-convulsive hemiplegia, inequality of the deep reflexes, unilateral abolition or impairment of the abdominal reflexes, and extensor plantar response. Eventually the appearance of hemiplegia, hemianesthesia, hemianopsia or aphasia with optic neuritis leaves no doubt of the diagnosis. Babinski's second sign is even more conclusive of organic origin of the fits. If the patient lies on his back with legs separated and arms crossed on the chest, and attempts to sit up, the paralysed leg rises with flexion of the trunk and thigh.

(8) *General paresis* gives no difficulty if the patient is examined properly. Dementia paralytica may cause convulsions indistinguishable from epileptic attacks or they may be unilateral with or without loss of consciousness and often followed by a transient hemiplegia. The delusions of grandeur, speech disturbances, tremors, handwriting, elision of syllables, lack of insight into their condition, gradually decreasing intelligence, ophthalmoma, etc., are quite characteristic. Juvenile paresis is also distinguished by the mental symptoms. The mental deterioration precedes the convulsions.

(9) *Brain Syphilis*.—This cannot always be separated from epilepsy as many cases of epilepsy are syphilitic in origin. The case should preferably be classed, however, as cerebral syphilis if there exist headache, transient paralysis of cranial nerves or inequality of the pupils. A positive blood Wassermann test, or an increase in the cells of the spinal fluid with increase of globulins and a positive Wassermann, occurs very frequently in epileptics without other symptoms than convulsions.

(10) *Uremia* may give rise to convulsions which are general or unilateral—or the same condition (Bright's) may give rise to localized edema of the brain with convulsions. The temperature may be elevated, but is usually subnormal, the convulsions follow the prolonged illness of the patient, the evidences of cardiovascular renal disease are present and the urinary findings are usually conclusive. The presence of hypertension, edema, albuminuric retinitis, etc., are valuable aids in diagnosis. Uremic, diabetic and acetoneuric convulsions belong to a different type from epilepsy. Contrary to some textbook statements, glycosuria does not follow epileptic attacks.

(11) *Malingering*.—The previous history is of value. The existence of a motive should excite suspicion. The choice of time and place, when being observed and never when alone, and only since the crime was committed, may lead to detection. There is rarely any injury in the fall, no scars, burns, or biting of the tongue. The urine and stools are not passed, but could be by a smart malingerer. The convulsion is usually a poor imitation and rarely does the malingerer think to put together all of the usual signs of a fit. They can simulate the

cyanosis but not the facial pallor at the beginning of the attack. The pupils are not dilated, and react well to light. There is no fever after the attacks. In epilepsy the thumb usually rests in the palms of the hands and is covered by the fingers.

(12) *Vertigo*.—Vertigo must be distinguished from minor attacks. Rarely does unconsciousness occur; there is no convulsion. Tinnitus aurium with prolonged giddiness and a clear consciousness are usual. There are often signs of internal ear disease. In Ménière's disease there are tinnitus aurium, sudden deafness, evidences of hemorrhage or inflammation in the labyrinth, nausea, vomiting, nystagmus, and the patient may fall unconscious. After the attack there may be a persistent irregular gait or staggering.

(13) *Febrile convulsions* occur especially in children, and often take the place of a chill in an adult.

(14) *Convulsions due to drugs* differ materially from epilepsy, and there is no history of previous attacks. In strychnin poisoning consciousness is retained. Convulsions may be produced by cocaine, arsenic, chloroform, ether, physostigmin, antipyrin, camphor, camphor monobromate, theophyllin, lumbar anesthesia with stovain, chronic lead poisoning.

(15) *Narcolepsy* must be differentiated from petit mal. Morbid somnolence, somnambulism, somnolentia (or sleep drunkenness) may be evidences of epilepsy. Epileptic automatism usually follows a fit (if it does occur) and more often a minor fit than a major fit, but may follow a major fit if the patient is aroused during the coma or sleep that follows the attack.

(16) In *tetany* and *tetanus* there is no loss of consciousness.

(17) *Hydrophobia* may resemble status epilepticus very closely, but there is no history of previous attacks.

(18) *Multiple sclerosis* presents marked physical evidences, such as nystagmus, scanning speech, intention tremor, etc., which cannot be confused with epilepsy.

(19) *Dementia præcox* shows the usual clinical picture of this disease.

(20) *Heart Block*.—In complete heart block the Stokes-Adams' syndrome may occur as a result of cerebral anemia and the auricle continues to beat, while the ventricle beats 5 or 6 times to the minute or not at all and epileptiform convulsions may occur.

(21) *Paramyoclonus epilepticus* presents shock-like contractions of parts of muscles, or whole muscles sometimes with and sometimes without loss of consciousness in the same patient. The movements are clonic and are not preceded by tonic spasm nor followed by coma, nor is the unconsciousness as certain or as profound as in true epilepsy.

(22) *Epilepsia* or *tuberous sclerosis* is characterized by numerous rounded elevated tumors of the cortex of the brain, associated with tumors of the other organs of the body and especially with adenoma sebaceum on the face, nose and mouth. The disease begins in the seventh month of fetal life, but the convulsions may begin in very early life or late childhood.

Complications.—**INJURIES.**—The patient may be injured in various ways. The fall may produce injuries when the patient strikes the floor or some sharp object, burns from falling against a stove or fireplace, or during the convulsions there may be bruises or injuries to bones or joints. Those working in certain occupations such as cab drivers, line-men, machinists, factory workers, etc., are especially liable to injury. By far the commonest injury is biting the tongue, which may be horribly chewed on the side on which the convulsions are stronger.

Some epileptics vomit after each attack. It may occur during the attack and lead to strangulation of the patient.

Scars are frequently present. Spratling found scars on the scalp and face in 238 out of 825 patients, and fractures in 29 of these cases, most frequently the nose and clavicle, occasionally the skull, legs, arms, or ribs. Burns occurred in 12 per cent. of the cases. In 22.5 per cent. there were scars on the tongue; occasionally there are dislocations of joints produced by the violent muscular contractions, such as the humerus or jaw. Mechanical purpura may occur. The teeth are sometimes broken by the clonic spasms of the jaw and the enamel may be worn off or chipped.

NERVOUS DISEASES.—The disease is not infrequently complicated by other nervous diseases, such as hysteria, neurasthenia, psychasthenia and mental conditions such as various psychoses, dementia, etc.

Soukhanoff has reported three cases of epilepsy associated with post-hemiplegic hemiathetosis.

Rupture of small blood-vessels in the skin and conjunctiva sometimes occurs. Whether the cases of cerebral hemorrhage starting with convulsions are cause or effect has not been proven, but it is certainly caused by the fit in some cases. Hematemesis occurs occasionally in epilepsy, according to Oster. Edema of the lungs occurs rarely in epileptic fits and may prove a fatal complication.

Sequelæ.—The most common and most important of the sequelæ of epilepsy is *mental deterioration*. This is usually in direct proportion to the frequency of the attacks. The patient first exhibits defective memory, irritability, a tendency to fight, is quick tempered, unreasonable, selfish, often lazy, with occasional periods of excitement. Sometimes explosive maniacal attacks replace the convulsions. Various psychoses are apt to develop and eventually a gradually increasing progressive dementia. Some cases develop mania, melancholia, paranoia or catatonia. Epileptic delirium occasionally occurs. Petit mal is more often associated with mental enfeeblement than grand mal, and the prognosis is worse. Severe convulsions in infancy are sometimes followed by arrest of moral development.

Epileptic insanity is characterized chiefly by confusion, incoherent speech, and unreasonable acts without any self-control, undressing in public, stealing, running away, urinating in a room. These attacks may last from a few minutes to an hour, rarely longer. Epileptics, as a rule, are inclined to be quarrelsome when conscious of their acts, are irritable, hard to please, hard to live with, willful, want their own way

about everything, until after many attacks mental enfeeblement sets in when their weaker wills bow to the wills of those about them. Sometimes they develop a delirium with violent maniacal excitement, a rapid flow of ideas, rapid speech, motor excitement, with hallucinations and delusions which may lead to acts of violence, destruction, arson, or other crimes. These attacks may last for hours or weeks with usually no memory of what has happened. In other cases there are anxiety, terrifying visions, stupor or apathy (Oppenheim). Eventually dementia occurs.

Association with Other Diseases.—Epilepsy may be associated with *migraine* and in some cases attacks of migraine replace the epileptic spells. Cases exist in which convulsions, migraine or vomiting attacks, one or the other, occur from time to time, suggesting that these symptoms are interchangeable phases of the same malady.

Ulrich claimed that 20 per cent. of cases of migraine were either directly or indirectly related to epileptic conditions. He concluded that migraine may be an epileptic equivalent; both may be present in the same subject. Migraine may finally merge into true epilepsy and epilepsy may replace migraine.

Labyrinthine symptoms and *vertigo* may be associated with epilepsy.

Petges reports a case of epilepsy following *typhoid fever*, and this is especially common following typhoid meningitis. Typhoid fever usually causes a cessation of epileptic attacks, as does any prolonged fever, but they return after the fever subsides. The patient may remain free from them for several months before recurrence.

Raynaud's disease is often associated with epileptic fits and probably represents vascular spasms or localized edema of the brain, similar to the phenomena occurring in other parts of the body.

Epilepsy is, of course, frequently associated with various *brain diseases*, such as tumor, meningitis, encephalitis, lead encephalopathy, cerebral syphilis, hydrocephalus, multiple sclerosis, the commonest being infantile hemiplegia. Epilepsy occurred in 41 out of 135 of Osler's cases of hemiplegia in children. Of all brain tumor subjects 60 per cent. have convulsions (Elsner).

Lombardo has reported three cases of *paramyoclonus* associated with epilepsy, and cases are also reported by Fazio in which paramyoclonus developed in epileptics. Advanced cases of *tabes* occasionally develop epileptiform attacks.

Osler quotes Salter as saying that *asthma* may alternate with epilepsy. Convulsions are often associated with any of the *acute infectious diseases* and with various *poisons* as lead, ergot, etc. Epilepsy is a rare complication of pericardial paracentesis.

This subject is further considered under etiology.

Epilepsy is associated with *heart disease* in 10 per cent. of the cases of epilepsy.

Falta states that Redlich has collected 72 cases in which epilepsy was associated with *tetany*.

Healy found that 7 per cent. of young *repeated offenders* were defi-

nitely epileptic, in which statistics he has not included infantile eclampsia or doubtful cases.

Shaw found the number of epileptics with *tuberculosis* 20.8 per cent., as compared with the average in England of 12.8 per cent.; and the tuberculosis death rate was 25.2 per cent., while the rate for England was 16.6 per cent. He found that 83.3 per cent. of epileptics reacted to tuberculin, against 33.3 per cent. of non-epileptics.

Between 8 and 9 per cent. of cases of *dementia præcox* have a history of convulsions in childhood (Ciese).

Clinical Varieties.—The clinical varieties are as follows:

Grand mal or *major attacks*, in which there is loss of consciousness with a convulsion.

Petit mal or *minor attacks*, in which there is a loss or impairment of consciousness with or (usually) without a fall, motor twitching, sensory auras, or other slight disturbances which may be the auras of major attacks.

Psychic epilepsy, in which there are transient mental disturbances in repeated attacks.

Jacksonian epilepsy, in which there is a localized spasm at times associated with a loss or impairment of consciousness, and at times developing into a general convulsion.

Symptomatic epilepsy, in which the epilepsy is a symptom of organic brain disease, such as tumor, abscess, trauma, infantile hemiplegia, hydrocephalus, localized cerebral edema, meningitis.

Uncinate fits, in which there are smell or taste sensations with smacking or chewing movements, associated with a dreamy confused state, and often a sense of fear.

Idiopathic epilepsy, in which there is no ascertainable cause for the attacks.

Myoclonic epilepsy or muscular jerking occurring in paramyoclonus multiplex on slight peripheral stimulation. The disease is familial and hereditary, and the myoclonus is at times associated with grand mal attacks. In half the cases the epilepsy appears before the myoclonus, and in a third they occur at about the same time (Austregesilo and Ayres).

Spastic spinal paralysis may exhibit violent clonic convulsions on the slightest touch—so-called *spinal epilepsy* (Osler).

Serial epilepsy, in which major or minor attacks occur close together in crops, or a series of attacks close together with a return of consciousness between attacks, and the patient is then free from them until another crop or series starts.

Status epilepticus, in which the spells occur one after another, so that the patient does not regain consciousness from one before another starts. This is usually accompanied by a rise in temperature, often by hyperpyrexia (105° to 107° F. [40.5° to 41.7° C.]). This condition may last for several hours or several days and always leads to great exhaustion and often to death. This condition may be brought about by a sudden cessation of treatment, as bromids, etc. Usually it occurs without

at an **apparent cause**. Equivalents of status epilepticus have been described such as hallucinatory confusion with fever or coma without convulsions. Some writers limit the use of the term "status epilepticus" to cases in which there is no return of consciousness between the spells.

Often the attacks start in one part of the body as a tonic spasm, which spreads in a definite order over the body to be replaced in the same sequence by clonic convulsions, and by the time the clonic convulsion has reached its "destination" the tonic spasm has again started at the point of origin.

Nocturnal epilepsy may remain unknown for many years if the patient sleeps alone, but may be suspected from bed-wetting, occasional periodic temporary headache, mental dullness and irritability on waking, lapses of memory, somnambulism, unexplainable outbursts of temper, finding the pillow wet from saliva, or the tongue bitten, etc.

Epilepsia procursiva, in which the patient runs before the actual attack.

Retropulsive epilepsy, in which the patient runs backward.

Poromania or *epileptic wandering*, in which the patient may travel or go some distance during a period of hours, days or weeks with apparently purposive acts, but awakens eventually with no knowledge of what has transpired. This is similar to the hysterical dual personality in which, however, the facts can be recalled from the subconscious mind by the usual methods. The truancy of mental degeneration (the moron class) is not associated with a loss of consciousness.

Senile epilepsy or *epilepsia tarda* occurs in old age and is associated with vascular changes (arteriosclerosis, arteriolar sclerosis, atheroma), and the attacks are frequently followed by paralysis and speech disturbances.

Epilepsia alternans has been described by DeVries in which the eyes and head are affected on one side and the extremities on the other side, the lesion being above the fifth nucleus in the tegmentum.

Epileptic equivalents, such as causeless attacks of perspiration with or without loss of consciousness, *narcolepsy* from which the patient cannot be awakened, or vasomotor disturbances, may occur. Many other symptoms are questioned by Oppenheim as being attributable to epilepsy, such as certain forms of angina pectoris, paroxysmal tachycardia, spasm of the glottis, asthma, neuralgia, transient hemiplegia, aphasia, profuse salivation, gastric disorders, *nocturnal enuresis*, transient deafness, priapism, paroxysmal bulimia or pica, general fatigue and apathy. It is a very safe guide not to accept symptoms as a part of epilepsy unless they are associated with a disturbance of consciousness at the time of their occurrence, or unless they immediately precede or follow an attack, or occur repeatedly in cases known to have epilepsy.

"*Epilepsie larvée*" or abortive forms (larval forms), in which there are repeated attacks affecting various body functions.

Muttering epilepsy, in which there are jabbering, muttering, the repetition of senseless words or incoherent unintelligible speech, mumbling, etc., which occur as a phase of petit mal.

Epilepsia continua is a condition in which muscular twitching occurs in different parts of the body between the convulsions and is usually due to an organic brain lesion.

Epilepsia nutans or nodding spasm may be a phenomenon of epilepsy.

Turner refers to attacks of a *vasomotor* character, throbbings, thumpings, beating of the heart, flushing of the face, fullness in the head, dizziness, paresthesia in the extremities, such as numbness, tingling and prickling, attacks of coldness of the limbs, pallor of the face, shivering, tingling and numbness, and sometimes slight tetanoid spasm.

Gowers has described *vasovagal attacks* as follows:

(1) Sensations referred to the stomach, heart and respiratory systems.

(2) The ascent of the sensation from the stomach to the chest, throat and head.

(3) The sensation is accompanied by a feeling of respiratory distress and cardiac oppression, fear, and a sense of impending death.

(4) There is no true loss of consciousness, but the mental operations are slow, and sometimes characterized by a feeling of unreality.

(5) The attack ends with a great acceleration of the heart's action.

(6) The whole attack lasts for about fifteen or twenty minutes.

Women are affected more often than men.

Syndrome of Unverricht—myoclonus and epilepsy.

Syndrome of Kojewnikow—partial continuous epilepsy.

Treatment.—**PROPHYLAXIS.**—The prevention of epilepsy must begin with a proper choice of a partner in marriage, and the avoidance of those who are neuropathic or belong to neuropathic families. The conversion of epileptics into asexual or anerotic individuals has been considered. Certainly those who are neuropathically tainted should avoid having children. An individual who is born of unhealthy stock should especially avoid alcohol, syphilis, and the many other etiological factors in the disease, and should give more than ordinary care to his general health.

GENERAL MANAGEMENT.—Attention should be given to the general health of the patient, such as **sleeping in the open air, work out of doors**, especially such occupations as gardening or farming, interesting vocations, recreations, such as golf, tennis, etc. They should avoid great mental strain and worry and should have a reasonable amount of sleep.

Frequent **bathing** and keeping the skin clean not only promotes elimination but lessens the probability of acne if bromids are given. If acne is troublesome, salt baths or salt fomentations are very valuable.

Farming and other outdoor work involving physical labor, fresh air and plain, wholesome food is the best occupation. Epileptics should avoid especially working around machinery, open fireplaces unless these are protected by fenders, avoid working around water, on telegraph poles, scaffolds and high buildings. Many cases should not be allowed to take a tub bath alone. They should avoid crossing crowded streets. Excessive mental or physical work involving strain should be avoided.

Confinement in an institution is valuable as a protection in very

severe cases who cannot be trusted alone, for feeble-minded patients, and those suffering from insanity or marked psychic attacks replacing or following convulsions.

Hydrotherapy is valuable in syphilitic cases, but does not offer any advantages in other classes of cases.

When convulsions occur at night, a hair pillow or no pillow at all should be used to avoid danger of suffocation. A daily **laxative** even if the bowels move every day is of value, and often a dose of **calomel** once or twice a week gives perceptible results.

DIET.—Writers differ greatly in their opinions in regard to diet. It may be stated as a general principle that the patient should eat three light meals daily, no food between meals, no excessive eating, little or no meat or meat derivatives (soup, extracts, etc.), no alcohol, no salt if bromids are being given. Foods that are hard to digest should be avoided especially by children. Vegetables, cereals, and milk are especially recommended. Fleury, also Rodiet, recommends a diet free from all animal foods, no milk, eggs, fish or meat.

All students of epilepsy agree upon the fact that there should be either a reduction or abstinence from meats. Nervous energy has its source chiefly in the **albuminous** and **nitrogenous foods**. Some allow meats only once a day, others not at all. A purin-free diet is recommended by others. Shuttleworth and Potts recommend little or no meats, limited sweets, no alcohol, no cucumbers or cabbage. Rosanoff in 15 cases reduced the proteid intake to one-half and found that for the seventy days of the experiment the spells were reduced in number by 14 per cent. On the other hand, Mackintosh reports a case in which he thought carbohydrate intolerance was the cause of the spells (boy, aged 4 years).

TREATMENT OF CAUSES.—It is always a safe proposition, in treating any patient, to correct everything that is found to be wrong with them. Especially is this true of diseases of unknown or uncertain causation, or in diseases where we know, as in epilepsy, that there are many different causes. A careful history, family, past and present, is presupposed, as well as a thorough physical examination. Sears on the head cannot be excluded without shaving the head of the patient. Certainly no case is complete unless the usual laboratory examinations are made, especially the blood and spinal fluid and stools. In the South, hookworm has been found a very frequent cause of epilepsy, and the attacks have been permanently cured by systematic and persistent treatment. *Tania saginata*, pinworms and *Ascaris lumbricoides* are very frequent in the history of cases of epilepsy, and when there is a history of these or when they are found, on examination, there can be no harm in using the appropriate treatment, though the results have not been so brilliant as with cases of hookworm.

Adhesions of the prepuce or clitoris are certainly part causes in some cases of epilepsy and the writer has seen cases recover without other treatment than the proper attention to these, though it must never be forgotten that when the disease is caused by these, not only the cause must

be removed, but the unstable condition of the brain must often be dealt with also; for the removal of a cause does not imply that the effect also has been removed, and an epileptogenic zone, when created by a peripheral lesion, must be rendered less sensitive by **sedatives** until the habit of convulsions has been broken up. When there is an **aura** beginning in a definite part of the body and a scar is found at this seat it should be removed. Any operation, no matter where or what, if done under a general **anesthetic** will usually cause a temporary cessation of convulsions, so we should be very slow in drawing conclusions from operations. Indeed, in some of the writer's cases the mere administration of an **anesthetic** without any operation has caused a suspension of attacks for several months.

Syphilis is one of the commonest causes of epilepsy. One of the writer's epileptic patients has a father and son who are also afflicted with epilepsy. She and her son both have positive Wassermanns, while her father died before she came under the author's care and the Wassermann test was not made. Such cases should have the usual antisyphilitic treatment. Even cases which give no positive evidences of syphilis often do better on **iodids** and **mercury** and **arsenic** than on any other drugs.

Alcoholic cases are treated like other cases of epilepsy, but, of course, **alcohol** should be discontinued, the bowels kept well open, and **bella-donna** with **tincture of hyoscyamus** given at frequent intervals.

TREATMENT DURING THE ATTACK.—When the *aura* consists of flexion of muscles, forcible extension will often prevent the attack. When the *aura* is a sensation in the leg or foot, the application of a tight **bandage** is helpful to keep off the attack. A **circular blister** has the same effect but of longer duration. **Rubbing** or biting the seat of the *aura* may stop the attack. Will power, running, or **muscular exertion** will keep off the attack in some cases. Any strong sensory stimulation has a similar effect, as smelling amyl nitrite or ammonia. The epigastric *aura* may be influenced by a **drink of whiskey** or **cold water**.

After the attack begins, the first effort should be to *prevent biting the tongue* which may be done by placing a hard rubber ring or pessary or a twisted towel between the teeth, which may be pried open with the handle of a spoon, or any other object may be used which will not cause injury to the tongue or teeth. One should never insert the finger between the teeth. Care should be taken to prevent the patient from injuring himself, and if time is allowed between the *aura* and the convulsive stage he should be placed on the bed, floor or soft ground. The collar should be loosened. It has been claimed that the attacks can be arrested or shortened by **placing the patient on his left side**, but some of the writer's patients claimed that this effect was produced by placing them on the right side, while the vast majority in which these were tried found that it made no difference on either side. Placing children in a **hot bath** or a bath to which **mustard** has been added, is often helpful and sometimes an enema will apparently shorten the attack. **Chloral hydrate** grains v to xxx (0.3 to 2.0 grams) or **sodium bromid** grains x to xxx (0.6 to 2.0 grams) may be given by rectum if the attack is prolonged.

Inhalation of chloroform until the convulsion ceases is by far the most certain remedy, but it must not be repeated at frequent intervals, as it will produce acute fatty degeneration of the liver. In status epilepticus it is especially valuable, and one of the writer's cases who had been having a spell every fifteen minutes day and night (100 a day) for several weeks did not have another for three months after administration of chloroform. In one case the writer chloroformed a patient once a week to prevent the spells, but after the fourth anesthetization the patient developed jaundice with enlargement of the liver. If vomiting occurs during the attack, the patient should be turned on his side and the mouth thoroughly cleaned.

In cases with auto-intoxication, **purgatives, diuretics and intestinal antiseptics** are recommended.

Treatment of Status Epilepticus.—By far the quickest and most potent treatment is **chloroform**, which, however, should be stopped as soon as the convulsions cease. Large doses of **bromids** or **chloral** may be given by rectum or the nasal tube. Bromids have been given hypodermatically, and also by lumbar puncture (sterile solution, grains xxx, or 2.0 grams, to the ounce) after withdrawing 10 or 15 c.c. of cerebrospinal fluid and injecting 10 c.c. of the bromid solution. The diet during status epilepticus should be liquid and may be given either by rectum or a nasal tube and may contain stimulants.

In the postconvulsive stupor stimulants are necessary, such as **strychnin, caffein, alcohol**, or if the pulse be rapid, **digitalis**. In acute dementia abundant **liquid diet, milk and eggs** should be given every two hours, and stimulants are also indicated. In acute mania restraint is necessary and **hyoscin** is often valuable.

TREATMENT BETWEEN ATTACKS.—**Bromids.**—By far the commonest drug used in epilepsy, and the one that most often gives good results is **potassium** or **sodium bromid**. The potassium bromid seems somewhat stronger than sodium bromid, but it also seems to give rise to more marked acne. There is great diversity of opinion as to the method of administration and dosage. Some writers prefer to give one large dose every night, while others prefer dividing it into three or four doses daily. No matter how young the child, the writer never saw any results with less than five grains three times daily. It has been his custom to give adults grains x (0.6 gram) three times daily—rarely grains xv (1.0 gram)—and if there is no effect from this some other drug is tried, either in combination with it or abandoning the bromid altogether.

Some competent physicians give as much as 180 to 300 grains (11.6 to 19.5 grams) a day while keeping the patient confined absolutely in bed, as it produces unsteadiness in walking and a drowsy, stuporous condition. To any one who has observed a large number of cases of bromid-poisoning, these doses seem quite unpardonable. The writer has seen bromid-poisoning occur from taking 240 grains (15.5 grams) in two days by mistake, and many cases in which it was produced by grains xx (1.3 grams) t.i.d. extending over a period of several weeks. When giving bromids, one should watch the tongue carefully and notice the

breath at frequent intervals, as these are usually the first signs of bromidism and, unless proper steps are taken, will soon be followed by a drunken unsteady gait, incoördination, disorientation, loss of the corneal reflex and stuporous insanity which often lasts from one to three months after the drug is discontinued.

Some writers think it advisable to give the drug in such doses as will produce toxic effects, as drowsiness, loss of the pharyngeal reflex, dilated and sluggish pupils, etc.; but it seems to the author, if it takes enough of the drug to produce toxic effects to ward off the attacks, it should be stopped and another drug substituted. The effect of the bromid is controlled largely by the amount of salt (common salt, NaCl) the patient gets. In order to get results from grains x (0.65 gram) t.i.d., the patient should be on a **salt-free diet**, otherwise the sodium chlorid will combine with the nerve cells and prevent the bromids from combining with them. Hoppe claims that one-third of the chlorin in the blood-serum has to be replaced by an equivalent amount of bromin before any therapeutic result is obtained, but when more than this is replaced bromid intoxication may occur. If the patient loses weight by abstinence from salt, it can be gradually increased to the point where it does not interfere with the bromids. When the attacks occur at a certain time of the day, it is well to time the bromid so as to have the patient under its influence at that time. Thus a patient of the writer never had any attacks except at six o'clock in the morning and was, therefore, required to take only one dose a day, which was at five o'clock. Patients who have their attacks only during the night should be required to take it on retiring or, if this does not stop them, give it on retiring and again four hours later, which can be easily managed with an alarm clock.

Patients sometimes complain very much of the *mental dullness* produced by bromids, in which case there is no objection to combining it with caffeine.

Acne is another frequent complaint, but can be easily overcome in most cases by giving **arsenic**, preferably in the form of **Fowler's solution**, or by local strong **salt water applications**, or, if general, by the use of **salt baths**.

The dose of bromids should never be larger than the smallest amount that will keep off the convulsions.

If a patient has *cystitis*, it will be made worse by bromids, or if he has ever had cystitis, it will probably return if bromids are given. But this can often be avoided by giving **hexamethylenamin** at the same time, especially if the urine is kept acid (by **acid sodium phosphate**). Any acute inflammation will be made worse by bromids and may necessitate its discontinuance. (Block, and Block and Nyun.)

The other forms of bromids have special indications. **Strontium bromid** requires about grains xx (1.3 grams) to obtain the same protective influence against the spells, as grains x (0.65 gram) of sodium bromid, but is valuable when the appetite is poor, as it seems to increase it. **Ammonium bromid** may be used in asthenic cases, or when the pulse is weak.

Muckenfuss gives the following quantities of bromin in the bromid preparations commonly used.

Potassium bromid	67.14 per cent.
Sodium bromid	77.64 per cent.
Ammonium bromid	81.57 per cent.

A combination of the three bromids is more effective than each alone. Theoretically the ammonium bromid should be the most efficacious form, but practically the author has not found this to be so. The **sodium bromid** has usually seemed most satisfactory.

Turner found that in 47.8 per cent. of cases, bromids had no influence or increased the frequency or severity of the spells, while in 23.5 per cent. they were arrested and in 28.7 per cent. were improved. He recommends from 45 to 90 grains (2.9 to 5.85 grams) per day. Laudenheimer (quoted by Turner, *Brit. Med. Jour.*, 1910) showed that an epileptic taking 150 grains (9.75 grams) of bromids a day for 8 days excreted a total of 35 grams (540 grains), or less than half the quantity taken.

As to the *length of time* a patient should take bromid, it should be continued for at least two years after the last convulsion, and *should never be stopped suddenly*, but gradually reduced a grain at a time, until after about six months it is left off entirely.

Bromids should always be given in a large quantity of water (6 ounces) and preferably after meals or at bedtime. It should never be given with alkalis, as they seem to destroy its effect. It is far more efficacious if given in an acid medium, such as digestive fluids containing hydrochloric acid.

Combination of Bromids with Sedatives and Stimulants. It is sometimes of value to prescribe a combination of bromids, thus: potassium, sodium and ammonium bromid grains v (0.3 gram) each, or in other cases to combine the bromids with other drugs such as **chloral hydrate**, **chloretone**, etc. In some patients who are prejudiced against bromids it may be used in the form of **sedabrol cubes** to make a cup of bouillon at each meal. Bromids are far more valuable in major than in minor epilepsy, but may fail in either.

Ethylene bromid has been recommended in 5-1,000 emulsion, giving 30 to 70 drops (1.9 to 4.4 c.c.) t.i.d. **Calcium bromid**, **gold bromid**, **zinc bromid** and many other forms of bromid have been recommended. **Bromipin** (potassium bromid with oil of sesame) (5 ss to 5 i, or 2.0 to 4.0 c.c.) may be used or it may be had in tablet form. **Bromipin** and **bromocoll** are said to produce very little acne.

The combination of bromids with other drugs is often valuable to enhance its effect or to overcome some of its unfavorable effects. Particularly is this true of the *heart stimulants*, such as **nux vomica**, **strychnin**, **digitalis**, **Tr. Adonis vernalis** and **cafein**. **Gélineau's dragées** contain **potassium bromid** 1 gram (grains xv), **picrotoxin** $\frac{1}{2}$ milligram (grain 1/200) and **antimony arsenate** $\frac{1}{2}$ milligram (grain 1/128) and is often very efficacious but hard to swallow. It is best to dissolve it in

water ($\frac{3}{4}$ vi). The **acid glycerophosphates** are often valuable in combination with bromids.

Clark recommends general **hygienic methods** in preference to sedative treatment by bromids, which he thinks often obscures the cause of the trouble. "I have seen many patients treated by sedatives steadily advance in physical, mental and moral deterioration in the face of a steady cessation of fits; suppression of fits is squared by an outburst of status epilepticus or a furious maniacal outbreak."

Spratling thinks that if recovery takes place under the use of bromids, it is in spite of, and not on account of, the drug.

Turner gives the following statistics on cures:

<i>Prebromid period</i>	<i>Postbromid period</i>
Hufeland 5 per cent.	Nothnagel 5 per cent.
Reynold 10 per cent.	Lachr 6 per cent.
Trousseau 13 per cent.	Ackerman 7.6 per cent.
Herpin 50 per cent. (?)	Dana 7.5 per cent.
	Wildermuth 8.5 per cent.
	Hoberman 10.3 per cent.
	Alt 12.5 per cent.

Other Drugs and Methods of Treatment.—Next to the bromids, the most efficacious drug in the writer's experience has been **Tr. simulo** 5 i to ii (4.0 to 8.0 c.c.) t.i.d. It should be given after meals. **Tr. belladonna** is sometimes valuable in major epilepsy, but is especially valuable in minor epilepsy. The dose, however, must be gradually increased as a tolerance seems to become established. It is best to start with minims v (0.3 c.c.) every four hours, and if not enough gradually increase to minims x (0.6 c.c.), using, of course, the smallest dose that will keep off the attacks. **Atropin** may also be used with similar results. **Borax** (grains v to xv, or 0.3 to 1.0 gram) combined with **nitroglycerin** (grain $\frac{1}{200}$ [$\frac{1}{3}$ mg.]) or **sodium nitrite** sometimes gives good results. **Antipyrin**, **opium**, **scopolamin**, **Radix artemisiae** have been recommended.

Digitalis was formerly in great favor in England for the treatment of epilepsy, but the writer has seen little or no results from its use. **Tr. Adonis vernalis** (minims x, or 0.6 c.c.) has often given good results. There is no objection to the use of strychnin. It never increases the frequency of the spells, and in asthenic cases often seems to do good in a general way. Iron, however, should never be given as it decidedly increases the frequency of attacks. **Camphor monobromate** (grains i to iii, or 0.06 to 0.2 gram) helps the giddiness in epilepsy.

Valerianate of zinc has some advocates, but the author has never seen any results from it. He has had no experience with **cannabis indica**.

[Recently the administration of **luminal**, a product now manufactured in America, has yielded excellent results in idiopathic epilepsy. It is administered in doses varying from $1\frac{1}{2}$ grains (0.1 gram) to twice that amount, given once or twice daily. In the milder cases one dose at night is usually sufficient. The drug is obtainable in tablet form, each

tablet containing $1\frac{1}{2}$ grains (0.1 gram), or the powdered drug may be dispensed in capsules.—Editor.]

Solanum carolinensis is one of the most valuable drugs in the treatment of epilepsy, and may be given in the form of fluid extract 5 i to iii (4.0 to 12.0 c.c.) t.i.d., either alone or in combination with bromids.

Sodium eosinate is of little value.

Chloretone is especially valuable, and particularly in nocturnal epilepsy, grains v (0.3 gram) being given on retiring.

Veronal often does good.

Calcium lactate, which is the only calcium salt used by the writer in epilepsy, seemed beneficial in some cases, while occasionally the patients were made worse. Donath found **calcium chlorid** of very little value. Perugia found no effect on the attacks by using calcium salts in 36 cases, though Littelljohn stopped the spells in 2 cases.

Chloral hydrate is a very valuable drug, especially in combination with bromids.

Dial is of some value, but causes giddiness.

Antipyrin grains v (0.3 gram) with **ammonium bromid** grains xv (1.0 gram) has been recommended by Andriezen (*Brit. Med. Jour.*, 1899) to promote mental brightness.

Paraldehyd.—Johnston recommends paraldehyd minims xx t.i.d.

Atropin.—Dorner recommends hypodermic injections of atropin sulphate, gram 0.003 to 0.006 (grain $1/20$ to grain $1/10$), in status epilepticus.

Borax is given in doses of grains v to xxx (0.3 to 2.0 grams) t.i.d., but it is apt to produce gastro-intestinal disturbances, or psoriasis. It is far more valuable when given in conjunction with nitroglycerin, grain $1/200$ (0.0003 gram) t.i.d. after meals, or it may be combined with bromids. Fowler's solution will usually prevent the psoriasis.

The **zinc salts** (oxid, valerianate, or lactate) are said to be occasionally helpful.

Alkalis have a beneficial effect, according to Shaw, and also oxalic acid and ammonium oxalate.

Strychnin is beneficial, especially in nocturnal epilepsy or when the blood-pressure is low.

Amylene hydrate, according to Oppenheim, has been recommended in aqueous solution (one to ten) in doses of 2.0 to 4.0 c.c. (5 ss to i).

Dormiol (30 to 45 drops a day) (2.0 to 3.0 c.c.) is said to do good.

Proponal (4.5 to 6.0 grams, or 70 to 90 grains, per day per rectum) has been used, but the writer has had no experience with it.

Opium, either in the form of morphin, codein or the fluid preparations, often stops convulsions, but it is of doubtful propriety for any chronic disease which is not to be fatal at an early date. By some opium is given for six weeks, alternating with bromids for six weeks.

Salvarsan is valuable in syphilitic cases, but in babies it is difficult and dangerous and they are very liable to choroiditis.

Bromalin (bromin and formaldehyd derivatives) has been used.

Bromocarpin (bromin and pilocarpin) is recommended by some.

Iron makes epileptics worse.

Arsenic is valuable and helpful.

Potassium or sodium iodid is valuable, even in cases which are not syphilitic, and will sometimes arrest the convulsions when bromids fail.

Mercury is certainly helpful in syphilitic cases and in cases with autointoxication.

Lithium carbonate, grains v to xv (0.3 to 1.0 gram) t.i.d. has been highly recommended.

Parnassia palustris with **paraldehyd** has been recommended. (Steep 3 ss [2.0 grams] of the herb in one pint of hot water for 15 minutes, strain, add 3 ss [2.0 c.c.] paraldehyd. Give one-third this amount t.i.d.)

Purgatives are very necessary in every case that shows intestinal putrefaction or absorption, and every case, no matter what the cause of the attacks may be, should be required to have a good bowel movement every day. Often a dose of calomel every week is of value with a saline every morning.

Serum therapy has sometimes been tried. Ceni's method of injecting the blood-serum of one epileptic into another has not given good results. According to Turner he "concluded that there existed in the blood of epileptics a soluble latent biochemical substance, an autocytotoxin, which has an influence over the elaboration of epileptogenic toxic agents and produces its results after repeated injections."

Crotalin (rattlesnake poison) given hypodermatically in doses of 1/150 to 1/100 grain every 3 or 4 days has seemed beneficial in some cases. Spangler found the severity and frequency of epileptic attacks influenced by injections of crotalin.

Bleeding and **lumbar puncture** have been tried in status epilepticus, occasionally with benefit.

Castia obtained marked benefit in 4 cases of status epilepticus by the withdrawal of 20 c.c. of cerebrospinal fluid.

Gordon withdrew 30 c.c. of cerebrospinal fluid from each of two epileptics and injected into the arm of each 3 c.c. of the other's fluid. He treated four cases in this way and in all there was a great improvement. In one case no cerebrospinal fluid was withdrawn.

Organotherapy has been used with various effects. **Thyroid gland** rather tends to increase the number of attacks, but sometimes helps the post-epileptic dementia. It is supposed to do good by overcoming intestinal stasis. Harrower thinks epilepsy due to thyroid deficiency in some cases. **Thymus gland** is also injurious and increases the frequency of attacks. **Orchitic substance** has been harmful in all cases in which the writer has tried it. **Ovarian extracts** make epileptics worse. **Pituitary substance**, especially the posterior lobe, is beneficial in the cases which have a deficiency. **Cerebrin** has no effect. **Suprarenal gland** is beneficial, but the writer never saw a cure result from it. It is theoretically supposed to do harm. **Parathyroid gland** grains ii (0.13 gram) t.i.d., increasing each dose by grains ii each month, and **calcium lactat** 3 i (4.0 c.c.) saturated solution daily are said by H. A. Knox to give

fairly good results. **Pancreatin** has been advised on account of the intestinal intoxication.

SURGICAL TREATMENT.—Any operation will usually cause a temporary cessation of fits if done under chloroform. We should, therefore, be slow to claim that a patient has been benefited by an operation until at least several months have passed. The chief value of operations is in traumatic and reflex epilepsy. The excision of a cicatrix, or freeing it from bone adhesions, sometimes gives good results. Especially is this true when the cicatrix is on the head, but is also sometimes of value when in other parts of the body, especially when the aura starts in the region of the cicatrix. When a nerve is buried in callus or fibrous adhesions, it should be freed, or nerve stretching may be practiced. There have been occasional reports of cessation of fits after appendectomy, operations for antrinitis, removal of polypi from the ear or nose, enucleation of a blind eye, but it is uncertain whether these cases were followed up for a sufficient length of time after the operations. Injuries to the skull, especially depressed fractures, foreign bodies, splinters of bones, adhesions of the meninges, cysts, or even gummata which fail to soften with iodids, offer fairly hopeful results. Horsley and others advise not only the removal of the bone and underlying pathological lesions, but also the underlying cortical center, to a depth of 5 mm., from which the fit originates. Sometimes we see cases of definite jacksonian attacks with no demonstrable lesion when the operation is performed, but even in these cases the superficial layer of the cortex concerned should be removed. Graf (*Arch. f. klin. Chir.*, Bd. 56), out of a series of 146 cases of traumatic jacksonian epilepsy, found recovery lasting for over 3 years in only 6.5 per cent. of those operated on.

Operations on the brain should never be undertaken unless there is some definite evidence of a brain or skull injury or focal brain disease, such as a tumor, abscess, cyst or foreign body, or unless there are paralytic symptoms associated with the convulsions, or very definitely localized and constant auræ.

Removal of the superior cervical sympathetic ganglion received much attention in the French journals in 1898, following the work of Chipault and others, but as time went on more and more cases were found to have recurrences of convulsions and the operation lost rapidly in popularity. It is probable that the anesthetic had more to do with the improvement than did the operation.

Osler reports a case of a testis in the inguinal canal, pressure upon which would cause a typical fit. Removal of the organ was followed by cure. Bahnmüller failed to influence a genuine epilepsy in a 3-year-old horse by castration.

Permanent partial compression of the common carotid arteries has been tried with the use of silver wires by Eastman, but the results were not satisfactory.

Nasal polypi and adenoids should be removed, whether they are considered the cause or not.

Aural polypi are said to have been removed with cure of epilepsy, but the writer has never seen it.

Adhesions of the prepuce or clitoris should be loosened or relieved by circumcision.

Removal of scars on the face, head, arms or legs sometimes give good results when the aura or beginning of the attack coincides with the scar. Davidson reports two cases of epilepsy cured by removal of one or both ovaries.

When status epilepticus is due to a localized meningitis serosa externa, trephining has sometimes given good results.

Cranicectomy is not justifiable in microcephalus, as the development of the skull merely follows the development of the brain which is arrested at the fifth month of uterine life. When signs of pressure exist, as in oxycephalus, or where there is premature ossification of the fontanelles, it may be tried.

Trephining is, of course, indicated in clear cases of jacksonian epilepsy, tumor, abscess, cyst or fracture.

Colon operations, short-circuiting and resection are practiced by some surgeons in the cases with colonic stasis, with a wide difference in opinions, but great danger to the patients.

Prognosis.—**RECOVERY.**—The general impression that epilepsy is incurable or rarely cured has been produced mainly by the fact that there have been too many "*one method*" forms of treatment adopted and by the fact that so few physicians make a systematic effort to find the cause of the disease. It must always be borne in mind that the disease is due to an unstable nervous system plus an exciting cause. The prognosis depends largely upon the intelligence of the physician who examines the case. The entire subject of etiology must be reviewed with each case.

The most frequent figure given is that 10 per cent. are cured, while other investigators state that 10 per cent. are cured by the bromid treatment alone. When all of the various causes are studied and the patients persist in carrying out the treatment and the doctors treat the cases properly, the writer has no doubt that over 25 per cent. of the cases are curable, and he thinks he has placed this figure entirely too low. It is a question if it is ever proper to speak of epilepsy as being "*cured*," owing to the fact that the attacks may recur at some future time, as in one case where they recurred after an interval of twenty-seven years. It is perhaps best to say that the disease has been "*arrested*." The question naturally arises as to *when the patient is to be regarded as cured or the disease arrested*. Some writers think that a cessation of fits for three years, others five, others ten years, is necessary to conclude that the patient is cured. The author feels that if, after a patient is treated for two years without the occurrence of a fit, the treatment is then gradually reduced and no fits occur for a year after treatment is stopped, the patient may be considered cured. It is not, however, claimed that the patient now has a perfectly stable nervous system and that he will never again be subject to any of the various functional

nervous diseases, for in reality he will be more subject to them than a normal person as long as he lives, and among these diseases he will be subject to epilepsy. The prognosis naturally depends much on the faithfulness of the patient in carrying out the general hygienic measures, diet and treatment.

Sometimes the disease terminates spontaneously without any apparent cause, even after the convulsions have lasted many years. The attacks often cease during pregnancy, typhoid fever and other protracted fevers, but usually return later, sometimes after an interval of some months. Jones reports a case of epilepsy which ceased during pneumonia, returned during convalescence, and then ceased entirely. According to Hoppe the cause of the cessation of attacks during fever may be the retention of bromids, the elimination of which is retarded during fever.

As to the probability of cure or arrest, it may be said that cases in which a definite cause can be found offer a much better prognosis than idiopathic cases.

Spratling and Turner state that *sex* has no influence on prognosis. It was formerly thought better in females. Cases which begin treatment soon after the onset of the disease are much more apt to recover than those which have lasted a long time. We naturally expect and usually get better results in cases in which the cause can be found and removed, but sometimes the damage already done is so great that the removal of the cause does not give much aid in the treatment.

Alcoholic epilepsy is very unfavorable as to cure.

Epilepsy due to *cerebral hemorrhage* offers a very bad prognosis, while cases due to *cerebral syphilis* usually do well. After the fits start, a *hemiplegic epileptic* deteriorates physically and mentally more rapidly than idiopathic epileptics and they never recover.

Quite contrary to reasonable thought, *inherited cases* of epilepsy offer a very favorable prospect of cure, except in cases in which *mental impairment* exists, in which cases the prognosis is bad in proportion to the amount of mental impairment. When the Binet-Simon test shows the retardation to be as much as three years, the cases are rarely arrested. Whether mental defects are congenital or acquired, the prognosis is very bad.

The prognosis is always better in cases that are *preceded by an aura*, than in those in which no aura occurs. Often the aura has a protective influence and aids the patient in avoiding the dangers of unconsciousness, the fall and the convulsion, as in driving an automobile. A patient who has always had aura cannot rely upon them for the future, as some attacks will occur without them. Petit mal is far more difficult to cure than grand mal, which may seem contradictory, as petit mal may consist only of such phenomena as occur as aura of grand mal. Nevertheless, *the more severe the convulsions*, the easier they are to cure, as a rule, provided the spells are not too frequent and have not produced mental impairment. According to Turner, 49 per cent. of

grand mal, 35 per cent. of combined grand and petit mal, and 26 per cent. of petit mal, are influenced by treatment.

The *longer the interval* between spells, the more favorable is the prognosis. The frequency of the spells is in direct proportion to the degree of mental impairment.

The sooner cases are placed under treatment the better the chance for recovery.

Cases in which the spells occur *only during sleep* or *only while awake* are easier to cure than cases which have spells both day and night.

Age.—According to Elsner, the prognosis for control is most favorable for the cases that begin before the age of ten years. According to Spratling, when epilepsy begins before ten years of age the prognosis is not as good as in cases which begin between fifteen and twenty years of age. From the age of twenty to thirty-five, cases of idiopathic epilepsy are difficult to cure, but cases due to discoverable causes offer a better outlook. Cases beginning at puberty are usually favorable. Certainly the cases beginning late in life, due to vascular changes, offer a very bad prognosis as regards a cure, but the convulsions are usually far apart.

The prognosis in *traumatic epilepsy* will depend largely on the individual case. In depressed fractures of the skull the spells may stop for several years after operation and then return.

When *sunstroke* produces epilepsy it is usually due to organic changes (meningo-encephalitis) and, according to Elsner, is not influenced by treatment.

Epilepsy due to meningitis, brain tumor, encephalitis and other *organic brain diseases* offer little or no hope for a cure. Many of the cases following fevers belong to this class, and it will be found usually that acute mental symptoms, delirium and headache occurred during the acute illness in the cases that are followed by convulsions. In all the cases of posttyphoid epilepsy seen by the writer, the history and condition of the patients indicated that it was due to typhoid meningitis.

Reflex epilepsy offers a good opportunity for a cure, but it must be remembered that after the habit of convulsions is once established the removal of the cause does not usually stop the convulsions unless further treatment is carried out.

Symptomatic epilepsy due to skull and brain injuries offers fairly good opportunity to the brain surgeon.

According to Moon, the prognosis as to the future moral and mental condition of children with convulsions does not appear to be any better when the first fit is associated with a reflex cause which can be removed than when it is of idiopathic origin.

MORTALITY.—The mortality in status epilepticus has been variously stated at from 28.5 per cent. to 50 per cent. When computed on the basis of status periods (instead of cases) Clark placed the mortality at 14 per cent. The probability of death in a case of status epilepticus is influenced by the frequency and severity of the spells to some extent,

but death may occur from prolonged coma with little or no convulsive movements. A prolonged very high temperature is an ill omen. Cases which last over 36 to 48 hours usually die.

The mortality statistics show that there were 2,476 deaths per 100,000 population in 1912, and 157 deaths from non-puerperal convulsions and 3,808 deaths from convulsions in infants, part of which were presumably of epileptic origin.

Of the epileptic deaths there were 1,460 males and 1,016 females. The largest number in a single year was in the first year of life; 8, 7, and 9 for two, three and four years of age (respectively); with 30 to 38 per year from fifteen to forty-four years, and a steady decline from that age upward.

The average age of death in epileptics is twenty-nine and one-half years. According to Spratling, "out of every 100 epileptics who die about 4 do so as the result of a single seizure, about 24 as the result of status epilepticus, about 24 as the result of some disease of the lungs, chiefly tuberculosis, about 12 as the result of some accident, including suffocation in bed, about 10 as the result of some organic disease of the heart, and about 26 from all other causes."

Thom and Southard found that 21.7 per cent. of their cases died of tuberculosis, the next most common cause being bronchopneumonia, and next pulmonary edema (the last 18 out of 205 autopsies), not counting the 72 cases which were merely said to have died from epilepsy. According to Osler, in children under ten years of age, 8.5 per cent. of deaths are due to *convulsions* (only partly epilepsy). Spratling quotes the U. S. Census Bureau as stating that 3 per cent. of deaths are due to epilepsy.

Mode of Death.—Death may be caused by *accidents*, from the falls, especially among people who have hazardous occupations, such as cab-drivers, linemen, carpenters, machinists, painters, etc., or may occur from falling down stairs, striking the head against some sharp or hard object, falling into the fire and being burned, or crossing crowded streets. According to Spratling, approximately 3 per cent. of the cases of epilepsy die from accidents due to it.

Vomiting during the attack may lead to *asphyxiation* from the vomitus getting in the larynx or trachea, or suffocation may occur from the patient turning on the face during an attack, thus interfering with breathing. Epileptics sometimes *drown* from going in swimming, or in a full bath tub. Some cases die from *suicide* and some are *killed* on account of their unreasonable and ungoverned temper. It is comparatively rare for a patient to die in a single attack. Death may occur from rupture of the heart, or paralysis of respiration, or hemorrhage from biting the tongue.

Heart disease occurs in 10 per cent. of epileptics, but is not the actual cause of death in this number.

As to FUNCTION.—Epilepsy places a decided restriction on the occupations open to the individual. He must naturally avoid any occupation in which his life would be endangered by a spell. If the spells are

far apart and there is no mental impairment, these cases often lead very useful and even highly successful lives. Cases with some mental impairment may be useful in minor occupations, but all cases of epilepsy are employed rather from sympathy than from a desire for their services, if normal laborers can be secured. (*See further, under Treatment, General Management.*)

Pathology.—There are no pathological lesions which would enable any one to decide from an autopsy alone, either by macroscopic or microscopic examination, that a patient had suffered from epilepsy. There are, however, many lesions which render such a clinical diagnosis believable.

Out of 205 autopsies on epileptics, Thom and Southard found 129 abnormal-looking brains. Of the remaining 76, 8 had leptomeningitis. After deducting all cases with mental symptoms, only 8 cases were left which might properly be accepted for a study of idiopathic epilepsy, and 4 of these had organic evidences, such as facial paralysis and leptomeningitis.

Redlich thinks the term "genuine epilepsy" should be abandoned and regards it as an organic cerebral disease, although the morbid anatomy is not yet understood.

THE BRAIN.—Very little to the point is known of the pathology of epilepsy. Of course, in organic brain diseases and jacksonian epilepsy the pathology is well known for these particular conditions, such as brain tumor, abscess, cysts, maldevelopment of the brain, porencephalia, cerebral hemorrhage, hydrocephalus, cerebral syphilis, arteriosclerosis, localized edema of the brain, traumata, fractures of the skull, meningitis, meningo-encephalitis, adhesions of the meninges, calcified echinococcus cysts, hypertrophy of the brain, agenesis cerebri leading to microcephaly.

There is no doubt that the primary seat of epilepsy is in the brain, which, however, may be influenced by conditions in other parts of the body. There is something either in the finer structure of the brain which has not yet been discovered or something in the altered physiological working of the brain which produces the attacks. The seat of these alterations differ in different cases and can frequently be surmised from the order of onset and the aura in many cases. In those cases which begin at once with a loss of consciousness and a general convulsion without any focal evidences, the seat is in one of the latent areas of the brain, usually the frontal lobe. Ziehen attributes the clonic convulsions to the cortex and the tonic spasm to subcortical centers.

The disappearance of convulsions after a capsular hemorrhage separating the cortex from the muscles, the loss of consciousness, the psychic disturbances, the mental condition of epileptics, and experimental work on the brain can leave no doubt that epileptic attacks have their origin in the cerebral cortex. Pollock calls attention to the fact that in decerebrated animals experimental convulsions are tonic only, never clonic.

The microscopical changes found in the brain have been considered

by some as the cause of epilepsy, while others believe them to be the effect. Certainly the more prolonged the case the greater are the pathological changes as far as sclerosis is concerned. There are no obvious pathological changes in the brain in recent or mild cases (except those due to gross pathological lesions or injuries), but in old severe cases there is atrophy and sclerosis of some part of the central nervous system in more than half the cases. The nerve cells show slight chromatolysis or even advanced degeneration. The nucleus may be excentric and globose. The blood-vessels are dilated and show atrophy of the walls with perivascular infiltration and punctiform hemorrhages. The lymph spaces are dilated and contain a foam-like exudate. Hyaline or granular masses are found in the lumen of the smaller vessels and capillaries. There is an increase of neuroglia, especially in the outer layer of the cortex and of the cornu ammonis. The meninges show slight fibroid thickening and moderate leukocytic infiltration. Rupture of the vessels with extravasation of red blood corpuscles and a delicate unorganized foam-like exudate are always present. The lesions are widely distributed. The cerebellar membranes are often more affected than the cerebral membranes. The regions most affected are the frontal and occipital, the cornu ammonis, and around the fissure of Rolando. These changes are probably the effect and not the cause of epilepsy (W. A. Turner).

Turner does not consider the Betz cells generally met with in epileptics as being either the cause or the result of the disease, but regards this as an immature form. In 70 to 80 per cent. of epileptics' brains some writers have found that the nerve-cells retain many of their infantile characteristics and persist among the fibers of the white matter (heterotopia).

Atrophy of nerve-cells in the brain has been reported, also proliferation of the neuroglia in compact bundles in the external layers. This sclerosis or gliosis may be visible macroscopically. Whether these changes are cause or effect has not yet been determined. Atrophy of the geniculate body and optic thalamus has been described by Onuf. Sclerosis of the cornu ammonis has been found in some cases.

Often slight adhesions of the meninges are pulled loose in removing the skull and are overlooked entirely. The writer has seen several cases of jacksonian type of epilepsy operated upon, with no history of head injury and only after the head was shaved for operation was a small scar on the scalp discovered. After trephining slight adhesions of the dura and arachnoid were found under these scars.

Thickening of the cranial bones, such as occurs in rickets, is sometimes found. Encephalitis, concussion, small hemorrhages, etc., may form the starting point of epilepsy. Hemiplegic idiots are often epileptic. Since many of the cases of epilepsy occur in feeble-minded and backward children we would naturally expect to find defective brain development present in many cases.

It may be stated generally that any organic disease or injury of the cortex of the brain is capable of producing convulsions. Certainly there

is abundant evidence that the chief seat of the disease is the cortex of the brain. Stimulation of an axon produces only one contraction of a muscle, while stimulation of the cell-body causes a series of contractions. According to Oppenheim, Ziehen attributes the clonic convulsions to the cortex and the tonic spasm to subcortical centers. While Binswanger and others thought the medulla oblongata particularly concerned in the production of convulsions.

Injuries to the spinal cord and involvement of nerves in cicatrices sometimes exist in epilepsy.

Krumholz reports a case of epilepsia continua due to localized encephalitis and gliomatous proliferation in the left motor cortical region. Lucas and Southard found convulsions in 7 out of 12 cases during encephalitis in children and 2 cases were followed by epilepsy.

Lapora found amyloid degeneration in the protoplasm of the nerve-cells in various parts of the brain and cord in a case of epilepsy which he considered due to an intoxication which also caused the myoclonic attacks.

Tilman found that in traumatic epilepsy the lesion was situated in the bone in 32 per cent. of the cases (mostly depression), in the dura (adhesions or thickening) in 9.3 per cent., in the arachnoid (edema, hemorrhagic infiltration, angiomas or cysts) in 38.4 per cent., the brain substance was involved (areas of softening or sclerosis) in 6.7 per cent., and no lesion was found in 13 per cent. According to Tilman, the injury may have occurred 10 or 20 years before the convulsions begin.

Sclerosis of the cornu ammonis occurs in about 50 per cent. of cases. In recent cases of pure epilepsy no sclerosis is found anywhere. There is as much reason to think the cortical sclerosis secondary as primary.

In status epilepticus Mott found great venous congestion and stasis and edema of the brain with arteriocapillary anemia, and distention of the perivascular lymph spaces.

Sioli found in a case of myoclonia and familial epilepsy a large mass of lipoid in the dentate nucleus of the cerebellum, invading the white substance, while in the case of Jacquin and Marchaud there were general cortical sclerosis, reaching as deeply as the pyramidal layer, atrophy of the pyramidal cells and degeneration of the tangential fibers.

THE HEART AND BLOOD-VESSELS.—Mott found extreme fatty degeneration of the heart in status epilepticus. Orbison reports a case of jacksonian epilepsy due to angioma racemosa of the pia. Abnormal narrowness of the aorta and cerebral vessels has been reported in some cases. Arteriosclerosis, arteriolar sclerosis and atheroma are of course common in the senile cases. In the cases of cerebral palsy beginning in childhood, micromelus of varying degree may occur.

THE BLOOD.—John Turner suggests that just before an attack, the leukocytes and blood plates shed their nucleoproteid; in consequence a hyaline material is found in the smaller arterioles and capillaries obstructing the free course of the circulation. These appearances are most conspicuous in the brains of persons dying in status epilepticus. W. A.

Turner suggests the probability that serial epilepsy and status epilepticus may be associated with nucleoproteid thrombosis arising from toxic influences within the body.

Ciard and Thom, in 160 blood cultures from 70 cases of epilepsy, found negative results in 156 cultures and contamination in 4 cases, and Wherry and Oliver had negative results in 6 cases.

There does not seem to be any sufficient evidence of the existence of Bra's neurococcus or Reed's epileptococcus.

CEREBROSPINAL FLUID.—Saniton and Chiray report a case of epilepsy two years after meningitis in which the attacks were followed by a transitory cerebrospinal leukocytosis and lymphocytosis.

Alfred Gordon states that there is a marked toxicity of the cerebrospinal fluid in epilepsy and that the accidental blood in the spinal fluid drawn from an epileptic underwent hemolysis which did not occur after the patient improved.

GLANDS OF INTERNAL SECRETION.—Claude and Schmieregeld found changes in the glands of internal secretion in epilepsy, namely, the thyroids, parathyroids, pituitary, suprarenals and ovaries, but there did not seem to be any characteristic findings.

According to Falta, parathyroprivia may give rise to convulsions, and he reports a case of tetany associated with epilepsy, with disturbances of the thyroid gland, and quotes Redlich who collected 72 cases of epilepsy associated with tetany.

Cushing reports 10 cases of hypopituitarism and uncinate gyrus seizures from neighborhood pressure, and other cases in which pituitary hypoplasia existed and he attributes the defect to the posterior lobe.

Thom found in a study of 42 cases of epilepsy that the relation of the weight of the liver to the brain was altered. Normally the liver-brain ratio is 7 to 6. In 62 per cent. (26 cases) the brain was found to be heavier than the liver.

History.—Epilepsy, from the Greek word meaning "to take or seize upon," a seizure, was originally spelled "epileny," and was known to the ancients as the disease of Hercules (who was supposed to have suffered from the disease), to the Romans as "morbus caducus," the "falling sickness." The Egyptians called the disease "morbus lunaticus et astralis," as they thought it due to the moon and stars. Among the Christians, the patron saint of the disease was St. John.

Hunt has written a very interesting sketch of the history of epilepsy.

Many names have been applied to the condition, the commonest being fits, spells, convulsions, spasms, the falling sickness, morbus sacer, morbus divinus. Spratling gives twenty-one different names which have been used for epilepsy.

Celsus first used the term "morbus major."

Bromids were used first by Sir Charles Locock in 1857.

Distribution.—Epilepsy occurs not only in man but many other animals, such as the horse, dog, and guinea pigs.

The disease seems to exist in all countries and among all races. Aside from North and South America and Europe it exists in Arabia,

Japan, Turkey, India, Australia. It is very common among negroes, and occurs also among American Indians.

Sociological Aspects.—The question of the marriage of epileptics is a matter of great importance. No absolute rule can be made, but each case must be advised on its own merits. It may be generally stated that only about 5 per cent. of the cases are fit to marry. The results of marriage must be considered from two standpoints:

- (1) The effect on the partner;
- (2) The effect on the offspring.

We can practically ignore the effect on the patient. Some authors state that marital relations make them worse, others that it makes them better. Thus Eulenburg states that sexual excitement either in courting or marital relations increases the frequency of attacks; Spratling found that, while it had no beneficial effect, it decreased the frequency and severity of the attacks for a time.

Of far greater importance than the effect on the patient is the effect on the partner. When we consider the disposition, character, and the attacks of patients (which are dealt with under Symptoms and need not be repeated here), we can readily see that they are not good partners to live with for a life-time. It is a great obligation and responsibility to nurse them and to feel that at any time a fatal accident may happen or that the disease may develop into the psychic equivalents, or actual insanity may occur. Antipathy, disgust, or fear is often excited in the partner and unhappy marriages and divorce or separation are not uncommon. Erotic impulses may lead to avoidance on the part of the other member of the family, especially when this may coexist with a quarrelsome, unreasonable and selfish personality. Women show more forbearance with their husbands than men show to their wives. In women, miscarriages, premature labor, or death of the child may occur from frequent attacks.

Of greatest importance is the effect on the progeny, who may have epilepsy or various other nervous diseases, such as hysteria, neurasthenia, migraine, imbecility, etc. Leuret found among 106 epileptics direct heredity in 11 cases. According to Féré, more than half of all the children springing from epileptic parents are subject to convulsions. According to Bouchet and Cazeauvieuille (*see* Senator-Kaminer), 37 from among 58 children of epileptic mothers died very young, and nearly all of them amid convulsions, while of the 21 who survived 7 more were suffering from convulsions.

Senator and Kaminer state that epilepsy is often associated with neuroses, neuropsychoses, exophthalmic goiter, diabetes (insipidus and mellitus), catalepsy, chorea, insanity, etc. (For further information *see* the subject of Heredity under Etiology.)

The probability of defective offspring is great enough to advise against marriage, or at least conception.

Naturally the exceptions to the rule would be the cases which have only very rare convulsions, far apart, and without mental impairment, and the cases with grand mal should be given preference over cases

with **petit mal**. Cases of epilepsy due to trauma are less liable to give rise to defective progeny than idiopathic cases, but even with these cases hereditary influence is probable.

Griffith says epileptics should never be allowed to marry, as they are usually prolific and their offspring are apt to be either mentally deficient, epileptic or both. Of his cases, 69.48 per cent. showed a family history of epilepsy, insanity, other nervous diseases, alcoholism or phthisis; 21.49 per cent. had a family history of epilepsy, while 40.18 per cent. showed a family history of phthisis. Only 45 out of 154 cases studied by him were females.

Davenport and Weeks find that epilepsy and feeble-mindedness show a great similarity of behavior in heredity, and that when both parents are either epileptic or feeble-minded, all their offspring are so likewise. When a case of migraine, chorea, paralysis or extreme nervousness is mated with a defective, about one-half of the offspring are defective.

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CHAPTER XXI

DYSKINESIÆ (Disorders of Motility)

By MOSES KESCHNER, M.D.

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TREMORS

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General Considerations.—The term tremor is applied to regular, involuntary, alternating movements taking place in smaller or larger excursions, always in the same plane, produced by involuntary contraction of certain muscles and their opponents.

A *simple tremor* is one which affects a single muscle group and its opponents. A *compound tremor* is one in which several groups of muscles and their opponents are in action and produce a complex movement, i.e., flexion and extension of the fingers combined with pronation and supination of the forearm. A *static tremor* is one which appears only during active movement when the involved limb is held in a special position. A *motor tremor* appears only during the act of movement.

An ordinary, voluntary, muscular movement is not the result of a continuous muscular contraction but consists of a "fusion" of rapidly succeeding, short, individual contractions from ten to twelve times per second; during fatigue or asthenia, the rate of these contractions be-

comes slower and less regular, the "fusion" becomes less perfect, and the result is a fine tremor.

Depending upon the nature of the disease which produces the tremor, it may appear only during rest, as in paralysis agitans, or during action (intention), as in multiple sclerosis, and in some cases only when the patient is under the influence of emotional excitement, such as pleasure or fear. Tremor may appear in paroxysms, as in hysteria, or may be present continuously, as in organic nervous disease or in toxic states.

In studying tremors, it is important to determine whether they are intensified or diminished during action or during rest, and what effect supporting the trembling limb has on their intensity; also what effect self-consciousness and emotion in general have on their intensity and amplitude.

Special consideration must also be given to the rapidity and regularity or rhythm of a tremor, so that we may distinguish between rapid and slow tremors. Tremors are considered rapid when there are from eight to ten oscillations per second, and slow when from three to five oscillations per second; there are also tremors which, in rapidity, stand midway between the two. Tremors may be fine or coarse; a rapid tremor is usually fine in its oscillations, and is known as a *vibratory tremor*.

When a tremor cannot be elicited during simple acts of movement, it may be elicited when the hand or leg of the patient is allowed to follow the examiner's finger while it is being moved away from it.

A tremor, arising from an exaggeration of the tendon reflexes or from increased muscle tone, is known as a *spastic tremor*, when it is elicited by the act of movement. *Nystagmus* is a variety of action or intention tremor of the eye muscles. A contraction and undulation which rapidly passes from one muscle bundle to another bundle of the same muscle, so that the whole muscle appears as if a wave passed over it, is a *fibrillary tremor*. This form of tremor is seen in definite pathological conditions of the nervous system, such as amyotrophic lateral sclerosis; it is aggravated by exposure to cold and during excitement, and may be seen in delicate and nervous individuals after cold, exposure of the skin, and physical overstrain, without the presence of organic nervous disease.

Transient quivering of a muscle affecting a few muscle bundles of one muscle without producing movement is called *myokymia*—popularly known as "live flesh," and is seen in anemic, weak, neurasthenic individuals. It is commonly seen in the orbicularis oculi, in the deltoid, biceps of the arm and in the glutei and quadriceps muscles of the leg. Myokymia is not associated with muscular atrophies, or with electrical changes, and is not affected by rest or voluntary exertion. It is of no serious significance.

Rhythmical variations, in the amplitude of tremors, have been observed with a certain regularity in each "tremor" movement, and such tremors have been designated as *allorhythmic tremors*. W. Salomonson¹ has found such tremors constantly in Basedow's disease, in poisoning with lead and mercury, paralysis agitans, alcoholism and hysteria

The variations, however, are very irregular, and have been found by most observers to have little or no relation to the movements of respiration and circulation. A regular allorhythmic tremor was found in paralysis agitans by Pollock,² and he ascribes the respiratory movements as responsible for the variation. Most neurologists attribute no diagnostic importance to the allorhythmic type^{3, 4} of tremor.

It is well to bear in mind that healthy people have tremors under certain conditions such as excitement, chills, violent physical exertion, and during convalescence from severe diseases. Pitres calls these tremors *physiological tremors*. Smoking and drinking to excess may produce a transient or persistent tremor.

Tremor in children, as an isolated or predominating symptom, except in the hereditary forms, such as hereditary nystagmus, is very rare. Sironi⁵ found only 3 cases of tremor during twenty-six years among the thousands of children at the Rome Children's Dispensary and 15 cases in the Naples Clinic. In children suffering from organic nervous disease, it is very commonly met with, especially as a symptom of chronic meningitis; it is also seen during the period of invasion of the exanthemata or other infectious diseases. Sironi cites a case in which the tremor in a child was due to the administration of thyroid extract.

Varieties.—**HABITUAL TREMOR.**—This form of tremor involves, most commonly, the head and hands, less frequently the face and tongue. It may be regular or irregular in rhythm. It is usually of slight amplitude, disappearing during rest, and intensified during voluntary action and mental excitement. Some individuals may be able to inhibit it temporarily by the exercise of the will. There is nothing definitely known of its *etiology*. Young individuals of a neuropathic tendency seem to be predisposed to it, and great physical hardships and mental shock, as fright or sorrow, are the exciting cause. The disease, as a rule, lasts throughout life; it may disappear spontaneously, and reappear temporarily after some new mental or physical shock. The *treatment* consists in attention to **general hygienic measures, hydrotherapy**, and the administration of preparations of **arsenic, belladonna** and the **bromids**.

FAMILIAL TREMOR OR "ESSENTIAL TREMOR."—This is a rare disease affecting individuals of neuropathic or psychopathic ancestry; there is usually a family history of chronic alcoholism, epilepsy, syphilis or insanity. Both sexes seem to be equally affected; it may be inherited from both sexes, several members of the same generation being affected.

This form of tremor has been observed at birth, but it may appear at any age; there are numerous cases in the literature in which it began earlier in life in each succeeding generation. Kreiss⁶ reports a tremor which affected several members of the same family in three successive generations, appearing earlier in life in each succeeding generation. The tremor in his cases involved every part of the body; it was mild during rest and became more intense during action. Mitchell has described a hereditary familial tremor affecting mainly the head. The most common involvement, in the order of frequency, are the hands, the leg and the muscles of the face and tongue.

The tremor is characterized by regular oscillations of small amplitude, from 3 to 9 times per second; it is diminished during rest, intensified on the execution of voluntary movements, and may be temporarily suppressed by the will. In some instances the tremor is not unlike that of multiple sclerosis. It is occasionally associated with typical choreiform movements, tic and nystagmus. (See Yawger's case of hereditary nystagmus of head and eyes in the chapter on The Myoclonias.)

The tremor may last throughout life, with periods of remission. Total disappearance almost never takes place, and treatment seems to have little or no influence.

SENILE TREMOR.—Old persons may present a tremor of the head and hands. The tremor is one of fine, rapid oscillations, increased by voluntary movement, diminished during rest and ceases entirely during sleep. It is distinguished from the tremor of paralysis agitans by the absence of rigidity and the absence of muscle weakness. It must also be differentiated from the tremor seen in exhaustion from prolonged and debilitating diseases. Senile tremor may be seen in individuals advanced in years, who show no signs of arteriosclerosis or organic disease.

TREMOR IN NEURASTHENIA.—In this condition the tremor is rapid, fine and vibratory in nature; it accompanies active movements and may be brought out by mental excitement. It is very similar to the tremor seen in healthy individuals after excessive smoking, drinking and sexual overindulgence. In addition to this form of general tremor, Oppenheim describes in neurasthenia a *fibrillary tremor* affecting the orbicularis palpebrarum, the orbicularis oris, the interossei, and the quadriceps femoris on exposure of the leg. This fibrillary tremor also has a tendency to appear during mental excitement or under the influence of cold, and may be so marked as to resemble *myokymia*.

TREMOR IN HYSTERIA.—The tremor characteristic of hysteria is one of large amplitude and of from 5 to 7 oscillations per second. It is intensified by mental excitement and voluntary movement, simulating the tremor of disseminated sclerosis in this respect, but differing from it in that it lasts longer than the movement which evokes it, or it may fail to appear in certain movements. It may be as marked during rest as during action; it is very indefinite and inconstant—*polymorphous* in type. It may appear in paroxysms, may be continuous, may be very slight or so marked as to develop into a clonic spasm. It may involve the legs so as to make locomotion impossible—or it may be most severe when the patient is on his back; it may affect any or all extremities or the entire body. Suggestion and hypnotism may diminish it or stop it.

TREMOR IN SHELL-SHOCK.—One of the commonly encountered symptoms in the neuroses which were observed during the last war, were the so-called "shell-shock" tremors. These tremors are coarse and irregular in rhythm, and may involve a part of the body or the entire body. Sometimes they are not unlike those of paralysis agitans; occasionally fine tremors, as seen in Basedow's disease, or intention tremors, similar to those in multiple sclerosis, may be seen. The slightest excitement tends to aggravate them; they are continuous during the wak-

ing hours, but cease entirely during sleep. A true functional tremor may be differentiated from a malingerer's tremor by ordering the patient to count slowly at first, and quickly afterward; if the rhythm of the tremor remains unchanged it may be considered functional.

TOXIC AND INFECTIOUS TREMORS.—Toxic and infectious conditions give rise to various form of tremor. The most commonly observed toxic tremors are those due to the excessive use of tobacco, coffee and tea. These tremors are usually fine and very rapid in character; they are exaggerated by exercise and mitigated by taking food. Those addicted to opium and morphin show a general tremor of the entire body, especially when deprived of their usual daily allowance of their habit-forming drug. Oppenheim once saw a tremor and lateropulsion in a gouty patient after colchicum poisoning, and made the diagnosis of paralysis agitans, but the symptoms promptly disappeared after the amount of colchicum was reduced.

TREMORS DUE TO CHRONIC METALLIC POISONING.—Chronic poisoning with metals such as bismuth, copper, arsenic, manganese, lead and mercury also give rise to tremor.

The tremor of mercury poisoning precedes the buccal symptoms and palsy, and involves the muscles of the face and extremities; it is usually constant, from 5 to 6 oscillations per second, coarse in nature, best elicited during movement, and increased during attempts at suppression; it is widespread and associated with great muscular weakness, general prostration, tremulous voice and mental deterioration. According to Noël Guéneau de Mussy,⁷ there are two distinct varieties of mercurial tremors. In one they simulate the shaking of paralysis agitans; in the other they are violent and occur independently of the will of the patient, and may be seen even when the patient is lying quietly in bed. On one occasion the author observed a man of thirty-seven, a mercurial thermometer filler in the service of Doctor Sachs in the neurological wards of Mount Sinai Hospital, whose chief complaint on admission was general tremor and prostration. The tremor had features both of multiple sclerosis and paresis; there were no signs clinically or biologically of either of these conditions. Treatment instituted for mercurial elimination was followed by a decided improvement in the tremor and the patient's general condition.

ALCOHOLIC TREMOR.—This form of tremor is more intense and coarser than any of the tremors observed in the neuroses. It is best seen in the extended fingers, lips and tongue; it is most marked in the morning before the patient has had his breakfast, and is diminished or may even entirely disappear after the administration of alcohol.

Quinquaud has described a sign which he considers pathognomonic of chronic alcoholism. When the examiner presses the palm of his own hand, which is held in a vertical position at right angles to the patient's fingers, which are fully extended at the interphalangeal joints, and widely spread apart, nothing in particular will be noticed for the first two or three seconds, but after this, the examiner will feel a slight quivering, jerking or crepitation, as if the phalanges of each

finger were knocking one against the other, trying to reach the examiner's palm. The same phenomenon may be elicited in tremors due to other conditions, and it is doubtful whether it can be considered pathognomonic of alcoholism. Alcoholic tremor is very marked during an attack of delirium tremens, and persists long after the mental symptoms of the condition have disappeared.

TREMOR OF BASEDOW'S DISEASE (Hyperthyroidism).—Although tremor does not always persist throughout the course of hyperthyroidism, it is, nevertheless, such a constant symptom as to be considered pathognomonic. It is a fine, regular, rapid and vibratory tremor, oscillating from 8 to 10 times per second (8 to 9, according to Kollaritz). It is most striking in the fingers and hands on extension, but may be seen and felt in the head, shoulders, trunk and feet; rarely is it limited to the trunk or extremities without involving the rest of the body. Irritation of any kind, cold, physical or mental excitement, aggravate the tremor, and although it is diminished during rest, it may in some cases be perceptible even then.

Tremulous respiration, in which the tremor is synchronous with that of the outstretched hands, is, according to Purves Stewart, one of the most constant signs of Basedow's disease.

The close relationship, which exists between the functions of the thyroid gland and the adrenals, has been utilized by Goetsch in the elaboration of a skin reaction known as the "*skin reaction of Goetsch*" which is diagnostic of hyperthyroidism. The reaction is obtained in the following manner: eight minims of a 1:1000 solution of epinephrin, diluted with an equal amount of clear sterile water, is injected hypodermatically into the patient's arm. Immediately an area of blanching appears around the injection, and about the margin of this area, usually, a red areola is visible, gradually shading off into the surrounding tissue. In about one-half hour the center of the white area becomes bluish-gray to lavender, and after about one and a half hours or two hours the red areola becomes bluish or lavender color, while that in the center disappears. The lavender areola remains for about four hours from the time of the injection, and is the most characteristic part of the reaction. Accompanying the local reaction, there is an exaggeration of the tremor and the nervous symptoms in general.

A general fine tremor is often seen in women during the menopause which is usually ascribed to hysteria or neurasthenia, but which will be found on careful observation to be due to thyroid dysfunction.

TREMOR IN PARESIS.—Tremor is a common and early symptom in paresis. It may be confined to some particular muscles or to one side of the body, or it may involve the entire body. It is most commonly seen in the lips and tongue; it is readily brought out by asking the patient to speak or to show his teeth, or to protrude the tongue. The latter movement is carried out in a very characteristic manner. The tongue is thrust out, showing a very fine and rapid tremor coming in waves along the muscles of the tongue, the mouth remaining wide open; or the tongue is repeatedly put out and drawn back.

The tremor of the fingers is also very common, but not as characteristic as that of the lips and tongue. It is fine and irregular, and may be slow or rapid; is intensified by physical or mental excitement, accompanies movement, and may persist during rest. The tremor in the extremities may be sufficiently severe to be one of the most conspicuous symptoms of the disease. Tremor in paresis may occasionally be complicated by the association with choreiform and myoclonic movements.

TREMOR IN INFECTIOUS DISEASES.—Tremor may be met with in the course of infectious diseases. Cases of poliomyelitis have been described in which tremor of the affected limb preceded the paralysis. Clement (cited by Oppenheim) saw a tremor, like that of paralysis agitans, with recovery in the course of typhoid fever. De Brun⁸ notes a tremor in every case of secondary malaria. The tremor, at times, is so intense that it interferes with work, writing, etc.; at other times it may be very slight. In two of his cases, the tremor was exaggerated by an impending malarial paroxysm. He believes that in some cases the tremor is toxic in nature, and in others that some organic lesion of the brain is responsible for it.

Various disorders of motility have been observed during the recent epidemic of "lethargic encephalitis." These may appear as *tremors* or *twitchings*, or both. In some cases they occur in the prodromal period and persist throughout the entire disease; in others they appear during its course. In either case they may remain in evidence months after all other symptoms have disappeared.

They may be localized in any part of the body, or may involve the entire body. The tremor may be fine or coarse, regular or irregular, rapid or slow; it may persist during sleep. Most of the tremors are typically parkinsonian in type.

In some of the cases generalized or localized *choreiform movements* were observed either alone or in combination with tremors, and in many cases the severity of the former overshadowed that of the latter.

The writer has seen a man of 50, who six months after an attack of lethargic encephalitis, showed choreiform movements in the muscles of the head, neck and both upper extremities; the movements were violent and jerky, and typical of Huntington's chorea. Another man of 35, at this writing—ten months after a severe attack of encephalitis—shows evidences of dystonia in both lower extremities. Neither case shows any other signs of the preëxisting encephalitis.

TREMOR IN ORGANIC BRAIN DISEASE.—Unilateral tremors, like those seen in paralysis agitans, are observed in *posthemiplegic motor irritations*, especially when the lesion involves the *lenticular nucleus*, so that pressure is exerted on the region of the cerebral peduncle and tegmentum. Holmes ascribes this tremor as due to cortical irritation which passes along the cerebellorubrospinal tract. *Midbrain* lesions may also give rise to tremors resembling multiple sclerosis. In these lesions the tremors are frequently associated with ataxia. A fine tremor has been observed in some cases of *frontal lobe* disease; the tremor then is confined to the upper limb on the same side as the lesion, and is best elicited when the

limb is held horizontally forward. Affections of the *cerebellum* or its *peduncles* also give rise to coarse tremors associated with *ataxia*, *dysmetria* (idea of distance), *asynergia* (incoördination), *adiadokocinesis* (failure to alternately extend and flex joints quickly and repeatedly, particularly supination and pronation) and *atonia* (absence of muscle tone).

CHRONIC PROGRESSIVE CEREBELLAR TREMOR (Hunt).⁹—From an analysis of three cases, Hunt concludes that there exists a chronic, progressive form of cerebellar tremor, the most striking characteristic symptom of which is a generalized volitional tremor which begins locally and gradually progresses. In its advanced stage, the disorder of motility is comparable in severity and violence with that of Huntington's chorea or generalized athetosis. There is, however, this difference, that in a position of rest and muscular relaxation the tremor movements cease. An analysis of the motor disorder shows a marked disturbance of the ability properly to control and regulate coördinated movements, as evidenced by the presence of hypermetria, dysmetria, adiadokocinesis, dyssynergia, hypotonia, and intermittent asthenia. All of these symptoms, including the volitional tremor, coincide with the classical symptomatology, which results from a loss of cerebellar control over voluntary movements. This disorder is, therefore, regarded as of cerebellar origin. The local onset, gradual progression, and chronic course indicate a progressive degeneration of certain special structures of the cerebellar mechanism presiding over the control and regulation of muscle movements. Other symptoms of cerebellar disease, such as disturbances of equilibrium, objective vertigo, nystagmus, cerebellar fits and seizures are absent. For this chronic, progressive disorder of the cerebellar mechanism, the name "*dyssynergia cerebellaris progressiva*" is suggested, as best indicating the essential element in the motor disturbance, dyssynergia, its progressive tendency and relation to the cerebellum.

Such cases have probably been variously classified as hereditary or essential tremor, multiple sclerosis, hysterical tremor, traumatic neurosis, and atypical paralysis agitans.

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THE CHOREAS

Synonyms.—Infectious chorea, Sydenham's chorea, Sydenham's disease, St. Vitus' dance, Chorea of childhood, Chorea minor, Danse de St. Guy, Danse de St. With, Myotyrbie (Dartigues), Periodical jactitation (R. Watt), Chorée, Veitstanz, Veitsdands, Vit-Tancz, Plasawicy seclotirbe, Corea, Folie musculaire (Bouillaud), Insanity of the muscles (Maudsley), Chorea Sancti Viti.

Introduction

The word "chorea," derived from the Greek (*χορεία*—dance), is suggestive of many different conditions which are all characterized by muscular movements, twitchings or spasms. The conditions in which such movements are the predominating symptom form a heterogeneous group, the component members of which must be clearly differentiated from each other. This group includes the infectious or Sydenham's chorea, with its various clinical types, the degenerative choreas (the Huntington group), the senile choreas, the choreas due to organic brain disease, the choreas due to the various forms of chronic poisoning, etc.

The most common affection belonging to this large group is infectious or Sydenham's chorea. In the discussion of this particular affection, the outstanding features of the other members of the group will be discussed, and the differences between them pointed out.

SYDENHAM'S CHOREA

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ings, p. 487—Pathology and pathogenesis, p. 504—Summary of etiology, pathology and pathogenesis, p. 506—Historical summary, p. 507—References, p. 508.

Definition.—Sydenham's or infectious chorea is a disease occurring chiefly in children, due to some toxic or infectious agent, which acts on the central nervous system by producing irregular involuntary contractions of the muscles, resulting in purposeless movements, and associated with muscle weakness and mental irritability.

Frequency.—Sée, in a hospital experience of 22 years, saw 531 cases of chorea; this number representing an approximate percentage of 0.8

per cent. of all his cases. Rufz found, among 32,976 sick children, 189 cases, about 0.5 per cent. Abt and Levinson found, among 10,150 sick children, 226 cases of chorea. Wicke observed the disease only in 0.18 per cent. of his cases. So many cases of mild chorea are discovered accidentally, that statistics as to frequency have little or no value. There is no doubt, however, that chorea is a common disease of childhood.

Etiology.—**PREDISPOSING CAUSES.**—*Seasonal Influences.*—Weir Mitchell, Sinkler, Sachs, and Starr found most of their cases to have begun during the spring. Gowers in his original cases found 33 per cent. occurred in the first three months of the year; 25 per cent. in the second three months; 20 per cent. in the third and 22 per cent. in the fourth. The greatest number of Abt and Levinson's cases occurred in January, the next highest in December, and the lowest in October. In the writer's experience most cases have occurred in the early spring months. According to McCarthy, of Philadelphia, the fact that most of the cases occur in the spring is most probably due to the poor general tone of the children after they have been confined indoors, and have been much occupied with their school work during the winter months.

Morris J. Lewis,¹ of Philadelphia, undertook a most elaborate inquiry, for a period of ten years, with a view of ascertaining whether there is any relation between temperature, humidity or barometric variations and the occurrence of chorea, but he could find none. He found that there is a slight correspondence with the average number of cloudy days, and also with the actual number of rainy days, and a still closer connection with the number of storm centers passing over Philadelphia; and that the wider the range of country included in these observations, the closer the correspondence, till an area of a radius of 400 miles was reached, and then the correspondence between the storm curve and the chorea became still closer. Comparing the average number of attacks per month of chorea and rheumatism, the curious fact was ascertained that the variations in the occurrence of rheumatism corresponded with those of chorea, but were uniformly a month later.

Sex.—The proportion of males to females is about 1:3. Starr found in 466 cases, 136 males and 330 females. The British Chorea Committee found in 436 cases 114 males and 322 females. Sinkler has collected 328 cases, of which 232 were females and 96 males. Gowers, who has combined the statistics of several other authors with his own, found that of 1000 cases, only 365 were boys. Sachs, in a successive series of 70 cases, seen within a year and a half, found 21 males and 49 females. In Osler's cases, the proportion was 1 male to 2 females. Abt and Levinson's cases were in the same proportion. In the last 30 successive cases seen by the writer, 11 were males and 19 females. According to Gowers the preponderance of girls is least in childhood, and increases after puberty; between 20 and 30 it is practically confined to females. This does not apply to the rare cases occurring during the second half of life, when they occur in equal numbers in both sexes.

Age.—Friedländer saw congenital chorea in two sisters. Both Mayo and Sinkler refer to a case of congenital chorea, due to a fright of the

mother during pregnancy. Richter, Fox, Heller and others also report congenital cases. Haven has seen 2 congenital cases among 195 cases of the disease. Simon has met the disease in children a few days old. Sachs claims to have seen several cases in children less than one year old; the youngest case of Wicke was 2 years old, and of Holt 4 years. Sinkler saw 2 cases in persons over 80 years. In Haven's² cases, 11 were under 6 years of age; 92 between 6 and 11; 58 between 11 and 15; 22 between 15 and 21, and 10 between 21 and 60. Gowers' statistics as to ages agree in the main with those of the British Chorea Committee, which are: 34 per cent. between 5 and 10 years; 43 per cent. between 10 and 15; 16 per cent. between 15 and 20; and 7 per cent. between 20 and 60. In Osler's 535 cases, 33 occurred in the first hemidecade; 228 in the second; 212 in the third, and 62 in the fourth. Abt and Levinson had the greatest number of patients between the ages of 5 and 14, with the highest percentage between 7 and 12.

It seems that the greatest number of cases occur between the ages of 7 and 13, the period of most active growth, when the greatest demands are made on the nervous system, and when metabolism is most active. It is doubtful whether the cases of chorea reported as first attacks (except in pregnancy) after the age of 20 are genuine Sydenham's chorea; in all probability those occurring after 40 are due to organic disease.

Racial and Climatic Influences.—Climate seems to have little influence on the disease. According to Axenfeld, on the authority of Rufz de Lavison, the disease is unknown in Martinique, Guadeloupe and other hot climates. It is comparatively infrequent in our Southern states. Weir Mitchell, of Philadelphia, Allen McLane Hamilton, of New York, S. A. Majure, of Dixon, Miss., and other busy practitioners report that they have never seen a case in a negro. Sachs claims to have seen, in the New York Polyclinic, a number of negro children with chorea, curiously all boys, in whom competition in school seems to have been the exciting cause. Osler, out of 175 cases of the disease seen in the Johns Hopkins Hospital, found 5 negro children among them; he found no case in a full-blooded Indian, but cases have been reported to have occurred in the half-breed. Germans and Americans seem to be more frequently affected than the Irish. It seems to be quite prevalent in Italy. Jews, with their natural tendency to nervous diseases, form a prolific source of the disease.

Hereditary Influences.—According to Burr,³ direct heredity plays no part in the causation of the disease. Less than 1 per cent. of the hundreds of patients with chorea treated at the Orthopedic Hospital and Infirmary for Nervous Diseases in Philadelphia, in the last thirty years, gave a history of the occurrence of the disease in either parent. The same author reports an instance known to him of 7 cases occurring in one family, in 3 generations, as an exception to the general opinion as to heredity, given in most textbooks. He considers heredity to be a greater factor in ties and habit spasms than in chorea. He believes the reason for the common opinion that nervous children are more prone

to chorea than others is the fact that all choreic children are nervous, fretful, peevish and emotionally unstable, and the physician is inclined to look upon this emotional instability as temperament, while in reality it is a symptom of chorea which quickly passes off during convalescence.

The literature is teeming with reports of cases similar to Burr's experience. The author has seen Sydenham's chorea occur at different periods, in three brothers, all of whom are subject to tonsillitis, and whose mother as a child had chorea and repeated attacks of tonsillitis and rheumatism. In the consideration of hereditary influences, the relation of chorea to rheumatism on the one hand, and to other nervous diseases on the other, must be borne in mind. An analysis of the cases seems to show that direct heredity in chorea is very rare, but that individuals whose ancestors have been afflicted with alcoholism, lues, saturnism, epilepsy, hysteria, ties, insanity or other nervous diseases (i.e., those with a tendency to neuropathy or psychopathy), are more pre-disposed to the disease than others.

Social Influences.—Poor children, especially those living in overcrowded, ill-ventilated tenements, and those who are poorly nourished, seem to be most susceptible. Bright and ambitious schoolchildren seem to be more frequently affected than those mentally retarded.

Imitation.—The disease is frequently brought on by "*imitation.*" It is doubtful whether the epidemics of chorea observed in boarding-schools can be considered genuine chorea; the muscular twitchings seen in these patients are most probably hysterical in nature. The writer has now under his observation two brothers, one a boy of five and one of seven years, both of whom seem to have true chorea. The disease began in the younger boy after an attack of tonsillitis, and four weeks later the older boy, without any apparent cause, developed typical chorea. There is a decidedly neuropathic taint in the family, and at the beginning of the twitchings in the older boy, we were inclined to attribute the movements to "*imitation,*" but the subsequent course of the disease leaves no room for doubt as to the exact nature of the condition.

Toxic Agents.—The grave cerebral types of *lead poisoning* are sometimes accompanied by choreiform movements of the limbs. Rubino reports a case of chronic chorea of 5 years' duration, in which chronic lead poisoning was the etiological factor; the choreiform movements were generalized, and persisted during sleep. The patient had also a typical intention tremor.

Chronic mercurial poisoning may be accompanied by an ataxia, a continuous shaking of the muscles of the face and extremities, with occasional ankle clonus. The movements interfere with locomotion, speech and mastication. Continued exposure produces amnesia, sleeplessness and exhaustion, and may terminate fatally. A peculiar brownish hue of the entire body, with a dry skin, generally accompanies the condition, which may give rise to tremors, typical choreiform movements, and muscle weakness, thus simulating chorea very closely.

Chronic poisoning with other metals may give rise to clinical pictures which, in a general way, simulate chorea.

The excessive use of tobacco has been observed in young adults to be the direct cause of "toxic" chorea. The twitchings are usually generalized, but they may be confined to the muscles of the face only; they are associated with vague paresthesias of the skin, cardiac palpitation, throbbing in the head, and gastric disturbances, and marked nausea, which is also a prominent feature. The slightest exertion brings on an attack of nausea very similar to the nausea of seasickness. Allen McLane Hamilton claims to have seen many cases of this disorder, and has found the feces often redolent of decomposed nicotine. The author has never seen a case of chorea, the origin of which could be traced to tobacco. An interesting case is reported by Rubino of a girl of seventeen employed in a tobacco factory, and who developed choreiform movements, which she was at times able to control; in addition to these choreiform movements she also had a tremor which was not, however, of the intention type. Because the chorea made its appearance after the girl's menses had been arrested, Rubino ascribes the disease partly as due to the "upset of the balance" in the glands of internal secretion.

Symptoms similar to ordinary chorea have been noticed during intoxication with hyoscin. In this condition, the choreiform movements are constantly associated with mild delusions. This symptom-complex is described in books as "*hyoscin chorea*." Poisoning with other drugs belonging to the vegetable group also gives rise occasionally to choreiform movements; but the symptoms are transitory and disappear promptly on the withdrawal of the drug. They can hardly be considered manifestations of chorea as described in this chapter.

Emotional Disturbances.—Fright, emotional excitement and general mental distress seem to play an important rôle as predisposing causes in the disease. All writers agree that if fright is to be considered a predisposing cause, the interval between the fright and the onset of the chorea must not exceed one week; the average interval is from 3 to 5 days. Most frequently, the chorea immediately follows the fright. Gowers cites the case of a boy in whom the movements began soon after a pistol had been unexpectedly discharged close to his ear; he also reports a case where a boy fell from an apple tree, after which he immediately began to shake, the tremor of fright assuming later a choreic character, and persisted as chorea. In children who have had chorea trivial occurrences, such as a severe thunderstorm or a severe scolding, may be sufficient to bring on a recurrence of the disease. *Overstudy* has undoubtedly much to do with the development of chorea, but the importance of *masturbation* as a cause has been overestimated.

Trauma.—Chorea may occasionally be traumatic in origin, but it is then dependent upon some grave cerebral affection or meningeal irritation. When it follows trauma without such an organic disturbance, it is most probably due to the coincident emotion.

Reflex Irritation.—Reflex irritation is believed to play an important part in the causation of the disease. Errors of diet—especially an extensive meat diet—are often said to be responsible for an attack of chorea; in these cases there is frequently a previous history of broken

sleep, night terrors, enuresis and somnambulism. Tardieu reports a case of chorea which was suddenly cured after the expulsion of eight *Ascaris lumbricoides*. Hanfield-Jones and Hamilton report similar cases.

Ocular defects, even low degrees of hypermetropic astigmatism, have been ascribed as causal agents of chorea. Stevens claimed that with the correction of the errors of refraction the choreiform movements had disappeared, but the New York Neurological Society after an impartial investigation of Stevens's claims came to the conclusion that the facts did not warrant their adoption.

Local spasms of the muscles of the face, the *habit chorea* of Mitchell, have been pointed out by Jacobi to be associated with enlarged tonsils and adenoid vegetations in the nasopharynx. These so-called habit choreas, however, are not, properly speaking, "choreas"; they are forms of facial spasms or ties.

Reflex irritations from the *genito-urinary tract* have, in the absence of any other cause, also been considered etiological factors in chorea. One of the most striking examples of the genital origin of chorea found in the literature is a case reported by Bossi⁴ in which, as soon as the endometritis and retroversion of the uterus were corrected, there was a prompt subsidence of the choreiform movements.

The fact that most cases of chorea occur at or near puberty, that it is more common in females than in males, and the peculiar course of the disease in the chorea of pregnancy are, in the opinion of some authors, arguments in favor of the *dysfunction of the glands of internal secretion* as being an etiological factor in the disease.

ASSOCIATED DISEASES.—In 1831, Thomson⁵ described "metastatic rheumatic inflammation of the cord and its nerves" as the chief cause of chorea. Bright,⁶ in 1838, considered post-rheumatic heart disease as a link between chorea and rheumatism. Hughes, in 1846, found, out of 104 cases of chorea, that 89 had either rheumatism or heart disease. Roger, in 1866, attempted to establish the fact that rheumatism, heart disease and chorea are parts of the same clinical entity, and he differentiated between "rheumatism-chorea," "cardiac chorea" and "cardiac rheumatic chorea," depending upon which one of these affections manifested itself. Jacobi, in 1875, in his essay on "Rheumatism in Children," pointed out the close relationship existing between rheumatism, chorea and endocarditis, and that usually *rheumatism* came first, later *endocarditis*, and last *chorea*. A reverse of this order he considered to be an exception and even then he had his doubts, because rheumatism in small children is often overlooked and considered to be "growing pains." Billiet was opposed to the views of Roger, Thomson, Hughes and Bright, because the coincidence of chorea with rheumatism is far below the frequency of rheumatism as a disease, and furthermore chorea affects females more frequently than males, while the opposite is true of rheumatism. Romberg could see no relation between chorea and rheumatism. Wunderlich believed the pains, so often manifested in chorea, to be an expression of hypersensitiveness and not of rheumatism. Steiner (1869) saw only 4 cases of articular rheumatism in 252 cases of chorea.

Gowers found a history of rheumatism in 25 per cent. of his cases. Osler found in 15.8 per cent. of his cases of chorea definite articular swellings, and in 5 per cent. of them rheumatic pains; 26 per cent. of Starr's cases gave a rheumatic history. Heubner describes chorea as an infectious disease, and regards it as much an evidence of rheumatism as gumma is of syphilis. Tylden reports in the St. Bartholomew's Hospital Reports a rheumatic family and personal history in 72 per cent. of his cases. In the 439 cases of the British Chorea Committee, 97 cases—or about 22 per cent.—gave a history of rheumatism. R. S. Eustis' found among 60 cases that 9—or 15 per cent.—were rheumatic on admission, and 19 more gave a history of previous rheumatism in the muscles and joints, making a total of 28 cases, of which 46.6 per cent. could be classed as rheumatic, and 13 cases, or 21.6 per cent., gave a history of both rheumatism and chorea. In the writer's last 30 consecutive cases, 11 gave a rheumatic history.

Macalister,⁸ in studying the life of the leukocyte, has shown that while the toxin in the blood plasma of chorea is toxic to the leukocytes of healthy persons, the blood plasma in cases of rheumatism is scarcely at all toxic, and that the plasma from chorea cases was toxic to the leukocytes of rheumatic cases; some authors conclude from this that the poisons in the two diseases are dissimilar. In spite of this, a study of the statistics of this subject justifies the conclusion that rheumatism bears a close relationship to chorea in about 25 per cent. of the cases.

Much speculation has been indulged in as to the nature of these relations. Some observers believe that emboli pass from the heart to the brain, and by occluding the small vessels, produce softenings which give rise to the choreiform movements. This cannot hold true, because, as will be pointed out under the pathology of chorea, emboli have only been found in a small number of cases, and then again, rheumatism may give rise to chorea without producing endocarditis. Others think that the infective process may cause thromboses in the small cerebral vessels. Pathological studies do not support this hypothesis.

With the advances made in recent years in bacteriological research, the attempt has been made to ascribe chorea as due to *bacterial invasion*.

Wassermann described a streptococcus, which he believed to be the cause of chorea. When injected into a guinea pig it caused a rise in temperature, swollen joints and choreiform movements. Dana found a coccus in a case of chorea with leptomeningitis of the brain and upper part of the spinal cord. Sachs has found a streptococcus in the blood of one of his patients with chorea. Meyer, Sander, and Cramer-Tobben also found streptococci in chorea. In a severe case of chorea and endocarditis, Triboulet found a bacillus. In two cases, Apert found Triboulet's coccus; Westphal found a staphylococcus in the blood and brain of a patient with severe chorea following articular rheumatism, and which, when introduced into the blood of animals, produced articular rheumatism.

Poynton and Paine cultivated the *Micrococcus rheumaticus* in cases of articular rheumatism, and they found the same organism in the brain

in cases of chorea, associated with rheumatism. Beaton and Walker found the same coccus in several cases of chorea. Camisa and Guevier have independently reported the finding of cocci similar to those of Poynton and Paine. Collins, in 1914, reported a case of chorea cured by the injection of an autogenous vaccine prepared from a coccus obtained by lumbar puncture.

Richards, of New York, found in the blood of two cases of chorea the *Streptococcus viridans*—an organism culturally similar to the green-producing streptococcus of Schottmueller, and to that found by Poynton and Paine. Both Hastings and Thro corroborated the identity of Richard's streptococcus with that of Schottmueller. It may be noted here, however, that in these 2 cases the cerebrospinal fluid was cultured and no microorganisms were found.

Dick and Rothstein⁹ isolated a streptococcus from the throat of a patient who had been suffering from chorea for 5 years, and injected it into a dog, which developed, 12 hours after the injection, typical choreiform movements.

H. W. Frink¹⁰ reports the case of a girl of 15 with chorea, from whose tonsils Thro isolated a pure growth of the *Streptococcus viridans*, similar to the *Micrococcus rheumaticus* of Beattie, an autogenous vaccine of which cured the girl's chorea.

Koplik¹¹ cultured the blood in many cases of chorea, but has never been able to find any microorganisms. Israel Strauss,¹² in 1915, cultured aërobically and non-aërobically the blood and spinal fluids of seven cases of chorea, but the *Streptococcus viridans* could not be demonstrated. He also inoculated monkeys intracranially with the spinal fluid of all the cases used. In one case, this had resulted in typical chorea for twenty-four hours, which then disappeared and returned for shorter periods. The monkey was killed and the brain was found normal; the heart muscle was examined for Aschoff bodies, which were looked upon as pathognomonic of rheumatism, or a disease, like rheumatism, in which an organism had not been obtained, but none were found. There had been two cases of chorea in which the heart showed Aschoff bodies. The individual, from whose spinal fluid the monkey developed chorea, died, and the autopsy showed encephalitis with hemiplegia. The cortex was removed through aspiration, and there had been hemorrhage. The material was then inoculated into monkeys but nothing developed from it. In the other cases the same procedure was repeated and nothing was found.

J. Donath¹³ made bacteriological examinations of the blood, cerebrospinal fluid and brain tissue in 7 cases of severe chorea. Two of the patients had amentia as a complication; 2 had chorea gravis which terminated fatally, and 2 of the uncomplicated cases had such severe twitchings that the blood had to be taken under narcosis. In 5 of the cases he found the *Staphylococcus albus*, and in 4 of these 5 cases the germ was isolated from the blood, and in the other, from the brain tissue. In the remaining 2 cases the *Staphylococcus pyogenes aureus* was obtained from the blood and spinal fluid, respectively. In one case,

Sarcina lutea was found in the brain tissue, and in another, in the blood. In some of the cases undifferentiated diplococci were also obtained from the blood. This author has no doubt of the infectious nature of the disease, but he does not think that these bacteria are the specific cause, but that they predispose feeble and anemic individuals to the disease.

The clinical fact that *tonsillitis*, *abscessed teeth*, *aural discharges*, and *other focal infections* have been followed by chorea which was promptly cured after the removal of these foci is considered as an argument in favor of the infectious nature of the disease. In a study of 1,000 tonsillectomies performed by Crowe, Watkins and Rothholz, they found that the removal of tonsils and adenoids is not a very satisfactory therapeutic or prophylactic measure in chorea; of the 23 cases of Sydenham's chorea, in which the tonsils and adenoids were removed by them, 8 had a recurrence of the chorea.

Helmholtz¹⁴ found, in 138 cases of chorea, 33 per cent. with a history of repeated tonsillitis, 21 per cent. of rheumatism, 26 per cent. of endocarditis, 8 per cent. of rheumatism and tonsillitis, 21 per cent. of rheumatism and endocarditis, and 54 per cent. of the entire group showed manifestations of one or the other of these diseases. Looking at the problem, from Rosenow's point of view, Helmholtz believes that the same organism which has a specific tendency to localize in the valves of the heart or in the joints may, under slightly different circumstances, localize in the brain cortex and produce chorea.

Quigley¹⁵ cultured the tonsils, blood and spinal fluid in 21 cases of acute, subacute and chronic chorea; 11 patients gave a history of acute tonsillitis; 2 of rheumatism, and one of endocarditis; 8 gave no history of either of these, and none had any evidence of lues. The blood was cultured in all the cases and the results were positive in 10 of them—9 of these gave small, slightly elongated cocci arranged in pairs, short chains and groups, and the tenth positive blood-culture, yielded a Gram-positive short diphtheroid organism. The spinal fluid of all the patients was cultured with thirteen positive results. Of these, 8 were organisms resembling those found in the blood. The throats of 15 of the patients were cultured, and 29 cultures were isolated and studied. The results showed 10 were hemolytic streptococci of variable size, occurring in long or short chains; 13 were green-producing cocci growing in pairs and short chains, and 6 were organisms which grew on blood agar as pinpoint, colorless, non-hemolytic colonies resembling the organisms from the blood in their morphologic and cultural characteristics.

Kinsella and Swift¹⁶ undertook to determine whether any constant cultural or immunologic type of bacterium was associated with acute rheumatic fever. They studied 58 cases and their conclusions are (1) that no type of streptococcus is constantly associated with acute rheumatic fever; (2) that the etiological relationship between the streptococcus and acute articular rheumatism cannot be definitely proven; (3) that, if the streptococcus is the etiological agent in acute rheumatic fever, it is through the various members of the viridans group, and hence no one member can be called the *Streptococcus rheumaticus*.

W. Lintz,¹⁷ from his researches on rheumatism, concludes that in some cases a microorganism can be isolated from the blood, but that the reason it cannot be found more frequently is because the bacteria tend to localize in the Aschoff nodules, and, except in the very virulent forms of the disease, are rapidly destroyed in the circulation. This investigator is not positive as to the exact nature of the organism, but he believes it to be similar to a streptococcus, and the reason why some animals fail to contract the disease after inoculation may be due to the lack of susceptibility on the part of the animal, or to the attenuation of the microorganism, or to both of these factors.

Morse and Floyd, from a study of 26 cases of chorea, are also of the opinion that a microorganism, or group of microorganisms, may be the cause of the disease, and that the source of infection is probably in the tonsils or teeth.

Reinhold reports a case of fatal chorea following sinus thrombosis, complicating pregnancy in a girl of 20. Stern collected in the literature 26 cases of chorea following sinus thrombosis; they were all fatal, but the results of the pathological examinations do not harmonize. Unless it was the same infection to which the thrombosis was due which produced the chorea, it is difficult to trace the relationship between the two conditions.

Many cases of cerebrospinal *syphilis* are accompanied by spasmodic muscle movements; these movements, however, are not typical of chorea. The older syphilographers described a form of chorea which they called "preparalytic chorea," and which was characterized by muscular spasms without the loss of consciousness, occurring before an attack of hemiplegia or paraplegia. When such spasms followed paralysis, they called it "postparalytic chorea." The twitchings were rarely general, and were usually confined to one arm and leg, or arm or leg alone, or face, arm and leg. These were probably choreiform movements due to *irritative luetic lesions* in the corresponding anatomical localizations, and were not what we would to-day call "chorea."

In recent years, by the great impetus given through modern biological and therapeutic methods to the study and investigation of syphilis, an attempt has been made to attach undue importance to syphilis as an etiological factor in chorea. Milian, in 1912, investigated the antecedents of a number of patients with chorea in various children's hospitals. He found in 11 out of 15 choreic children a positive Wassermann reaction; in 2 other children the syphilitic origin of the chorea was doubtful, but they presented stigmata commonly attributed to hereditary lues. He then announced his belief that he regarded syphilis, hereditary or acquired, as a most important etiological factor in chorea. Inasmuch as he was able to cure these cases very quickly with mercury, and because arsenic has been used empirically for years in chorea with good results, and is used with so much satisfaction to-day, in the form of salvarsan, in the treatment of syphilis, he considered these facts to be strong links in the chain of evidence in the support of his theory.

Numerous objections were raised to Milian's claim. Comby¹⁸ could

find evidences of syphilis in only 7 out of his 39 cases of chorea; 24 cases in his series gave a positive tuberculin reaction; 6 a history of acute articular rheumatism; and 8 had signs of cardiac involvement.

Guillain's objections were that the spinal fluids in patients with chorea show only slight meningeal involvement, there being very little, if any, lymphocytosis or excess of albumin. His cases, on autopsy, never showed the customary lesions of hereditary lues in the brain or cord. The histories of the cases of chorea, which he had seen while superintendent of the Salpêtrière, were always negative as to lues.

Guillain had employed salvarsan in the treatment of chorea by rectum (not intravenously) in doses of 0.1 gram, according to the method recommended by Weill, Mouriquand, Goyet and Morel; in 7 cases out of 10 the results were failures.

Triboulet found, in 400 choreics, only 4 with signs of hereditary lues. Babonneix found, out of 145 choreics, 36 in whom hereditary lues might be said to exist. Pierre Marie treated 25 patients with this disease by intravenous injections of salvarsan. The injections were administered on an average four times a week, the doses varying from 0.2 to 0.3 gram and even 0.35 gram. A cure generally followed after the third or fourth injection, so that the duration of the disease was reduced to three or four weeks. In spite of these favorable results, Marie does not believe that chorea can be considered to be of syphilitic origin.

Nobicourt and Tixier had never been able to discover any signs of lues in their cases of chorea. Chauffard is of the opinion that the good results obtained by the arsenical preparations in these cases are not due to their effect as specifics for syphilis, but rather as agents of general application.

Germanns Flatau¹⁶ reports a case of chorea of 5 years' duration with a history of hereditary lues, convulsions, meningeal symptoms and a positive Wassermann reaction, in which the ordinary treatment for chorea had no effect, but the administration of salvarsan and mercury was followed by a prompt cure. It is doubtful whether this was a case of Sydenham's chorea. Salinger²⁰ records a case of chorea minor in a girl of 10, with a history of hereditary lues, in whom after three weeks of treatment with salvarsan the choreic movements were entirely arrested. Szametz reports a case of chorea treated successfully by salvarsan, in a rheumatic boy in whom the chorea followed an attack of rheumatism.

Koplik²¹ found in 11 successive cases of chorea admitted to the Mt. Sinai Hospital, between the ages of 6 and 13, that 5 had endocarditis on admission, and in 3 there was a definite history of rheumatism or rheumatic pains. Of these, 10 cases were examined for the presence of the Wassermann reaction; in 8 it was negative, and in 2 it failed, but it was not positive. In none of the cases were there any stigmata of hereditary or acquired lues, nor could lues be traced in any way in their family histories. Nevertheless, 9 of these cases were injected with neosalvarsan in doses ranging from 0.15 to 0.4 gram; in 7 of the cases thus injected, no striking results were observed. The average duration

of the disease in these 7 cases was 36 days after the last injection; in one case a severe nephritis set in, and the patient was ill for some weeks after, although, eventually, recovery took place. Koplik concludes that "in salvarsan we have no agent of any value above what has hitherto been in vogue in the treatment of chorea minor," nor does he feel that we are warranted in tracing any relationship between Sydenham's chorea and hereditary or acquired syphilis.

Ilaberman²² reports the case of a girl 9 years old, with a positive Wassermann, and a history of difficult birth, deafness of four years' duration, scaphoid scapula, and a soft systolic murmur at the apex, best heard in the recumbent posture; the parents were both luetic. For the last three years the little girl had chorea which did not respond to any form of treatment, but was promptly and completely cured by vigorous antiluetic treatment. The same author also reports the case of a boy whose father contracted lues three years before marriage, and whose mother aborted three times and remained sterile for four years, when she finally gave birth to the patient. The boy developed well physically, but was a very poor scholar; he had great difficulty in concentrating his attention; he was active, but easily frightened; a restless sleeper and a poor eater. At the age of 6 he developed chorea which was not amenable to any of the ordinary therapeutic measures, but was promptly cured after he was subjected to antiluetic treatment.

At the Montefiore Home and Hospital, the last 14 successive cases admitted for chorea were carefully studied from this point of view, and in all of them the blood and cerebrospinal fluid showed no evidences of lues, nor were there any signs of hereditary or acquired lues. Veeder and Jeans found that 43 per cent. of their cases of late, hereditary lues had some form of involvement of the central nervous system, and of 32 patients with such involvement, there were only 2 cases of chorea. Morse and Floyd²³ found in their series of 26 cases of chorea that syphilis played no direct part in the etiology of this disease.

A critical survey of these statistics seems to show that syphilis might be an accidental, but is not a direct, etiological factor in chorea. A child affected with lues might contract chorea just as readily as any other child, and that the same holds true whether the lues is acquired or hereditary. Nevertheless, when a case of chorea has lasted over three or four months, and does not seem to progress favorably under routine treatment, it is well to bear in mind that lues might be at the basis of the disease.

The influence of *infectious fevers* on the development of chorea is important. Chorea may develop after *scarlet fever* at periods varying from six weeks to six months, after the attack of scarlet has subsided. Priestley²⁴ found 13 cases of chorea following 5,355 cases of scarlet fever—a proportion of 1 in 412. In Carslaw's 533 cases of scarlet, only 3 were followed by chorea. In the British Choreia Committee's report, scarlet fever is given as the sole antecedent cause of chorea in 6 per cent. of all the cases. Osler states that scarlet fever with arthritic manifestations may be a direct antecedent of chorea; about 25 per cent. of his cases of chorea gave a previous history of this disease.

Sturges (cited by Osler) states that a history of previous *whooping cough* occurs more frequently in choreic than in other children. Osler's infirmity records do not bear out Sturges' findings.

Measles as an antecedent to chorea is recorded in the British Chorea Committee's report in 26 per cent. of the cases, and in 7 per cent. it was the *sole* antecedent illness. Chorea may follow influenza, tuberculosis, typhoid, gonorrhea, small-pox, chicken-pox, diphtheria, cerebrospinal meningitis, pyemia or any infectious disease, especially when accompanied with hyperpyrexia and severe constitutional disturbances. The severity of the chorea bears no relation to the severity of the antecedent infectious disease. It seems doubtful, however, whether any of these conditions are to be considered direct etiological factors.

Kinnicutt, Heinemann and others have pointed out a relationship between chorea and certain states of malarial poisoning. In southern climates, where the cerebral forms of pernicious malaria, due to the estivo-autumnal parasite, are common, motor symptoms, varying from slight muscular twitchings to convulsions or typical chorea, occur frequently, but these never seem to appear in the first paroxysm. It is doubtful, however, whether these cases can be considered genuine choreas.

Symptomatology.—**CLINICAL HISTORY.**—In most of the cases the temperature remains normal throughout the disease. In severe cases, especially those due to or following infections, it may be 100°–102° F. (37.8°–38.9° C.). Uncomplicated fatal cases have been recorded with a hyperpyrexia (106°–108° F. [41.1°–42.2° C.]). R. E. Hare has shown that monochorea is associated with a rise of temperature in the affected limb. A difference of 4 or 5 degrees was often found to exist between the two sides of the body. Asthenia, loss of weight and exhaustion due to insomnia, and inability to take sufficient food are common in the severe cases.

PHYSICAL FINDINGS.—*Choreic Movements.*—The most characteristic symptom of the disease is the rapid, coarse, involuntary, spontaneous, irregular and purposeless movements. They are irregular in time as well as in character; they cannot be arrested for any length of time, and are increased by attracting the patient's attention, by excitement and by an effort to restrain them, or to carry out any volitional movement; they are ordinarily diminished during physical and mental rest and cease during natural or induced sleep.

At the beginning of the disease only occasional twitchings of the face or hand are noticeable, but as the disease progresses they become more frequent and more marked, and finally continuous. They may become so violent that the muscles of the entire body appear as if in constant motion.

Owing to the abruptness, rapidity and irregularity of the movements, work requiring exact coördination, such as writing, sewing, playing the piano or walking, cannot be performed; coördinated movements of the fingers, such as apposition of the thumb and index or little finger, are usually impossible.

Oppenheim has drawn attention to the tendency that these patients

have—in addition to the irregular and uncontrollable choreiform movements—to purposeless associated movements. Foerster has shown that in chorea the orderly coöperation of the agonists with the antagonists during voluntary movement is lost, but that single muscles or muscle groups participate in the contraction, e.g., in extension of the finger, the extensor communis digitorum acts without the interossei; in closing the fist, the wrist is not extended, etc. There is, in other words, a typical ataxia. A choreic patient, on account of this ataxia, never performs the same movement twice in the same manner.

Weakness of Muscle Power.—The next most common symptom is weakness of muscle power. This weakness may bear no relation to the severity of the choreic movements, and it may be considerable in extent before the latter are recognized, but there is never anything like a complete paralysis. This muscle weakness is called “choreic pseudoparesis,” and in many cases it overshadows and inhibits the movements. Close observation will show that the little patient uses one limb less than the other, or perhaps ceases to use it at all. He can still move the limb when ordered to do so, but he does so only for a short time and very feebly. The muscles become limp and hypotonic; the tendon reflexes are diminished, and in some cases even absent. As the disease advances the twitchings become more marked, and the pseudoparesis assumes secondary importance as a symptom. West called this condition “chorea mollis,” and Gowers²⁵ “chorea paralytica.”

While any muscle of the body may be affected by the choreiform movements, they are more noticeable in the extremities and face than in the muscles of the trunk. The involvement may from the outset be general, but as a rule is confined to one limb, or to one side of the body, but becomes generalized as the disease is progressing. In at least half the cases both sides of the body are not equally involved. The arms are almost always affected earlier and more severely than the legs; the facial muscles are very frequently affected, resulting in peculiar facial contortions and grimacing; the eyes are suddenly closed and opened, the mouth pouts, the tongue rolls within the mouth, and is pressed between the cheeks, so that speech becomes quick and indistinct. The words are shot out or jerked out in separate parts, the last syllables being entirely cut off. Laryngoscopic examination discloses an irregularity in the movements of the vocal cords. Involvement of the diaphragm results in irregular and “catchy” respiration, and further interferes with speech. According to Glogau,²⁶ the curve taken from thoracic breathing during speech will show choreic jerks, which cannot be demonstrated in other muscles.

Swift²⁷ of Boston, from a study of the voice in 20 cases of chorea, has found that there is a change of voice which is more frequent in the vowels, less so in whisper, whistle, consonants, air-blow and holding of breath—the frequency being in the order cited. He found sufficient uniformity and frequency in the appearance of vocal changes to warrant a classification of changes in pitch and intensity of the voice, as one of the signs of chorea. The most marked change occurred in the open.

prolonged sound of "a" as in "are." When the pseudoparesis involves the muscles of speech and respiration, the voice may be reduced to a whisper, and in severe cases the patient may become temporarily mute.

The involvement of the *tongue and mouth* may also extend to the pharynx and seriously interfere with the taking of food.

Involvement of the *muscles of the neck* causes frequent movements of the head to one side, the eyes moving with the head. The hands are alternately flexed and extended, the fingers are spread apart, the arms rotated inwards, and the shoulders are drawn up. Slight affection of the *trunk muscles* causes the patient to sway from side to side, when sitting or standing, but a more severe involvement results in a total inability to sit or stand. The limbs may be thrown about so violently that the patient cannot rest in bed, and frequently sustains severe contusions all over the body. Tuckwell records a case where spasm of the *muscles of the jaw* was so violent that several teeth were broken. *Involvement of the legs* results in their being thrown about violently, now extended; now pressed against each other, then again rotated outward, or abducted.

As a result of unequal spasm of the muscles of the *eyeballs*, temporary diplopia may be present. The pupils are dilated but react to light and accommodation. Von Ziemssen observed on several occasions the pupillary reactions to be sluggish. Hippus (spontaneous, rapid and spasmodic variation in the size of the pupil) has been noticed. Ophthalmoscopic examination shows the fundi to be normal. Embolism, atrophy of the disk and optic neuritis have been seen during or after attacks of chorea, but these changes are not characteristic of the disease. Concentric restriction of the visual fields has also been noted in chorea.

Electrical Irritability of the Muscles.—Uncomplicated cases of chorea do not show any alteration in the electrical irritability of the muscles. These retain their size, but in "paralytic" cases they are hypotonic. Elloy claims to have seen muscular atrophy in the involved muscles.

Reflexes.—The tendon reflexes are, as a rule, normal; they may, however, be increased or diminished. Joffroy has seen cases where they were completely abolished. Oddo believes the reflexes are normal in mild cases, and diminished or suppressed in severe cases. In the writer's experience the reflexes have either been normal or increased, except in paralytic cases, when they were diminished, but never absent.

Gordon and Eshner have observed cases in which, when the patient is on his back and a short blow is struck over the patellar tendon, the response will be like in the ordinary knee-jerk, but instead of the leg coming down immediately after, it will remain suspended in the air for some time and come down gradually, i.e., the knee-jerk has the character of a tonic muscular contraction. Oppenheim considers this phenomenon to be a coincidence, or that it may possibly be due to a reflex choreic contraction of the quadriceps tendon occurring simultaneously with the reflex movement. This may also be an explanation of the "wobble knee-jerk," described by Swift of Boston, as characteristic of chorea.

Sphincters.—The sphincters are not involved, except in the terminal

stages of the very severe cases, and in the cases with marked mental symptoms, as a result not of paralysis but of mental apathy. Enuresis in children, however, is quite common both at the beginning and throughout the disease.

Sensation.—Chorea is a painless disease. The incessant movements cause fatigue but no actual pain. When the disease is due to or complicated with rheumatism, there may be pains in the muscles, joints and nerves, but then the pains are not limited to the limbs affected with the choreiform movements. Chorea due to organic brain disease, with involvement of the thalamus and lemniscus, is very painful. In ordinary chorea the presence of hemianesthesia or paresthesia is usually indicative of a complicating hysteria.

Tremor.—A fine, regular tremor involving the tongue, lips and fingers may be observed in addition to the choreiform movements. This tremor is intensified after physical and mental excitement, and is probably due to the accompanying general debility.

Convulsions.—Convulsive attacks do not occur in chorea, except as manifestations of an associated hysteria or epilepsy. Gowers states that he has found in many cases of epilepsy that the convulsions made their first appearance after an attack of chorea.

Heart.—The most common symptoms referable to the heart are those due to mitral insufficiency. The organic nature of the murmur in this condition must be differentiated from the usual systolic murmur heard over the mitral or pulmonic areas, due to secondary anemia or conditions other than endocarditis. The latter is often associated with a venous hum in the jugular vein, and the murmur itself is not transmitted. The cardiac dullness, which may extend slightly to the right, may also be due to the anemia, and subsequently disappear. The endocarditis associated with chorea is usually of a mild nature, and may disappear without leaving any trace. Occasionally a faint systolic murmur may be heard at the apex with some beats and not with others, said to be due to irregular contraction of the papillary muscle. Cases complicated with rheumatism may show a friction rub over the precordial area due to a complicating pericarditis.

Pulse.—The pulse is usually increased in frequency, about 10–15 beats per minute, and on account of the irregular breathing the heart's action may also be irregular. Irregularity, however, is not as marked a feature in chorea as rapidity. The subjects of the disease being usually neurotic children, slight emotional disturbance is sufficient to produce a rapid, irregular and tumultuous heart action.

Blood-Pressure.—Owing to the accompanying anemia, there is a tendency to a somewhat lower blood-pressure than normal.

Skin.—Vasomotor disturbances, such as hyperidrosis, dermatographia, flushing of the face and neck, and general erythema are seen in children, with a tendency to spasmophilia and to ductless gland disturbances. Fissured lips, with herpes labialis due to irritation by the continuous smacking of the lips, are perhaps the most common symptoms referable to the skin and mucous membranes. Other cutaneous distur-

ances when present are probably due to poisoning with arsenic taken for therapeutic purposes. These are characterized by the presence of papular or erythematous rashes combined with pigmentation of the skin. Osler found two cases of herpes zoster in his series at the Philadelphia Infirmary. The various eruptions of purpura are not uncommon in the "rheumatic" cases. English clinicians have described subcutaneous fibrous nodules in chorea, but no such cases have been reported in this country. Children, with the so-called exudative diathesis, are predisposed to chorea, and, inasmuch as such children are also frequently affected with eczema, the latter has been considered a complication or manifestation of the disease. Some writers have even gone so far as to claim that a child, suffering from eczema, is most apt to become choreic when the eruption of eczema has been allowed to "strike in." It is doubtful whether any of the cutaneous manifestations seen in chorea are more than mere coincidences.

Rheumatism.—Rheumatism is so commonly seen in chorea that some writers consider it a symptom of the disease. In our opinion, it is a cause and complication of chorea, and is discussed in the sections on etiology and complications.

PSYCHIC STATES.—Abnormal psychic states play an important rôle in the symptomatology of this disease. The patients are irritable, peevish and forgetful; they are easily frightened, cannot concentrate their attention, are disobedient, and very fretful. Some of them have night terrors, and transitory auditory and visual hallucinations. Psychic symptoms, other than irritability, fretfulness and disobedience, are not as common and not as marked in children as in adults. All sorts of transitions, from the slightest to the most pronounced depressions, and typical manic depressive states with hallucinatory delirium, and even acute mania, have been met with. In adults the psychic symptoms are not unlike those of the infectious or intoxication deliria. The intellect, as a rule, does not suffer; if anything, the patient shows greater mental keenness. The mental symptoms are at their maximum at the height of the disease, and may persist after recovery. Severe mental symptoms make the prognosis much graver, because they may be the direct cause of the patient's death, or the patient may recover from the chorea and remain permanently demented; fortunately such severe mental cases are quite rare. Cases of chorea have been recorded in which the movements were insignificant as compared with the psychic disturbances.

LABORATORY FINDINGS.—*Cerebrospinal Fluid.*—In ordinary cases the cerebrospinal fluid is normal in quantity, clear and sterile, with no increase in cells. In severe cases it may be increased in quantity, and under high pressure. Morse and Floyd²⁸ examined the cerebrospinal fluid in 26 children with chorea: 11 boys and 15 girls. In one of the cases the disease was fatal, and several of them had a very severe type of the disease; in the remaining cases the course was mild or moderate. Lumbar puncture was done 20 times in 19 cases. The fluid was perfectly clear in every instance. In one case there was an apparent increase in pressure. It was normal in all the others. A fibrin clot was never

formed. The number of cells was counted in 10 cases and was respectively: 2, 5, 7, 8, 10, 10, 10, 18, 24 and 25. They were all mononuclear in every case. In a few cases of chorea in which the bacterial origin of the disease can be traced to some localized or general infection, bacteria may be found on culturing the fluid.

Blood.—In the majority of cases there is a pronounced anemia of the chlorotic type, with little change in the number of erythrocytes, or their shape, but with a low hemoglobin index. Leopold²⁹ of Philadelphia found in 20 cases of chorea an eosinophilia varying from 4 to 16 per cent. F. H. Leavitt³⁰ examined the blood in 80 cases, principally to determine the frequency of eosinophilia and its relation to herpes labialis. All were cases of Sydenham's chorea in the active stage of the disease, between the ages of 3 and 30, the average being about 10 years old; there were 53 females and 27 males; 79 were white and one was a full-blooded negro boy. He concludes: (1) That herpes labialis is most likely due to mechanical irritation by the patient himself, and that eosinophilia, when present, is due to the condition of the skin, and is not inherent to the disease, chorea, itself. (2) There is a great increase in the lymphocyte and a relative decrease in the polymorphonuclear leukocyte count. (3) A fairly constant low-grade leukocytosis is present in most cases, indicating the infective origin of chorea, rather than the theory of a functional neurosis. (4) That the anemia is of a chlorotic type, with little change in the number and character of the erythrocytes, but with a low color index.

Urine.—The urine is abundant in amount, and when the chorea is due to rheumatism, large amounts of urates, uric acid and phosphates are in evidence. During the height of the disease the specific gravity may be as high as 1.030 or even 1.035. Changes in the urea nitrogen output, albumin and casts, in the absence of cardiorenal complications, are very rare. Temporary glycosuria has been observed. Garrod found urohematoporphyrin in 14 out of 20 cases of chorea, and Herter found the same product, not only in chorea, but also in rheumatism. Choreic children frequently pass a turbid urine with a copious white precipitate of calcium carbonate and phosphates; the total elimination as a rule is not increased, because the usual proportion in the stools is lower, to balance the increase in the urine. These findings are not characteristic of chorea. Kleinschmidt and others have found similar contents in the urine in children with hysteria, epilepsy and general neuropathy. Provinciali found in a girl of 14 with chorea, that she retained only 0.918 gram of calcium instead of over 30 grams, and that nearly 4 grams were eliminated in the urine and feces.

Duration.—The average duration of the disease is from six weeks to six months, but it may last only three weeks or longer than six months. The average duration of the cases collected by the British Chorea Committee was about ten weeks. All of the cases seen at the Montefiore Home and Hospital, which is an institution for the treatment of chronic diseases only, have been of no less than three months' duration, a good many six months', and a considerable number more than one year: two

cases had the disease for more than two years without interruption, prior to admission. There are cases recorded (Mildner⁸¹ and Macdougall⁸²) in which the disease began in youth and lasted till death, which occurred in one case at the age of 66 and in another at 50.

As a rule, the severer the disease the longer is its duration. The age and sex of the patient, the condition of the heart or the other complications do not seem to bear any relationship to the duration of the disease. Slight choreiform movements, the so-called "residual chorea" (Guthrie), may persist for months.

Recurrences.—Recurrences in chorea are so common that they are considered a characteristic feature of the disease. More than one-third of Gowers' cases had recurrences. In Bing's cases more than 25 per cent. had recurrences. Although Gowers reports cases in which recurrences took place at the same season in several successive years, the prevailing opinion seems to be that seasonal recurrence of the disease is not common. Two or more recurrences are quite common. Sachs and Peterson found that, out of 70 cases, 18 had a second attack, 11 had a third, 4 had a fourth, 1 had a fifth, and 1 had a sixth attack.

Females are more liable to recurrences than males. The disease seldom recurs between the ages of 18 and 30, except in pregnancy. After the disease has once ceased, the slightest cause, such as fright, overstrain, change of climate or season, or an acute illness, may bring on a recurrence. Contrary to what would be expected, preceding rheumatic fever seems to have no influence on the occurrence of a relapse. Gowers found in his cases a history of rheumatic fever in precisely the same proportion of the recurring cases as in the whole series. Organic disease of the heart and pericarditis occur more commonly during a recurrence than during a first attack.

The intervals between the relapses, except in pregnancy, vary from a few weeks to one or two years; in chorea complicating pregnancy, there may be an interval of ten years from the last attack to the recurrence. The intervals between subsequent attacks (recurrences) present great variations; they may vary in the same patient, and there is no uniformity as to the time of the year in which a relapse may occur, as compared with the previous attacks.

A relapse may imitate the first attack, as regards its origin and distribution, but there is no definite rule about this, as there is none about the severity of the relapse as compared with the severity of any of the previous attacks.

Diagnosis.—The disease is easily recognized by the peculiar movements which can be elicited in the mildest case by making the patient hold both hands above his head; after a few seconds, especially when his attention is distracted, rapid, irregular twitchings appear in the fingers of one or both hands. Another good method of bringing out the movements—particularly in the mild, unilateral cases—is to let the patient grasp the examiner's hands; the difference between the uniform muscular contractions of the healthy side and the unsteadiness of the grasp on the affected side will then be readily noticed. In the

cases in which the legs are more involved than the arms, the station and gait may simulate paraparesis, but the twitchings will be sufficiently evident to show the exact nature of the trouble.

DIFFERENTIAL DIAGNOSIS.—*Paralytic Chorea*—*Unilateral Multiple Sclerosis*.—In paralytic chorea, when loss of muscle power is a predominating feature, diagnostic difficulties may arise. Such cases may be mistaken for genuine paralyses. In “paralytic chorea” the weakness is, in most cases, confined to one arm; it does not involve the face or leg, and on careful examination choreic movements will be observed; these, with the history of the gradual loss of the use of the affected extremity, will clear up the diagnosis. Grinker, of Chicago, showed a case before the Chicago Neurological Society on October 21, 1909, which he had seen three years before that, and had made the diagnosis of paralytic chorea; at that time the patient began to show clumsiness on one side of the body, and later, irregular choreiform movements began to make their appearance on the same side; there were no other signs. As time went on the patient developed a scanning speech with all the other classical signs of a unilateral multiple sclerosis.

Tic.—In distinguishing chorea from tic it must be borne in mind that in tic the movements are rapid, coördinate and purposive; they may be tonic or clonic, with comparatively long periods of rest between the movements. Tic is usually confined to certain definite parts of the body, most commonly the face and shoulders, rarely to the trunk or legs. A choreic does not repeat the same movement as regularly and as systematically as a tiqueur. No matter how violent a tic may be, it never interferes with voluntary movement. Tiqueurs are usually psychically abnormal.

Spasm.—Chorea is differentiated from spasm by the fact that in the latter the movements are very brusque and involve muscles corresponding to the anatomic distribution of a certain nerve. They are usually due to some reflex irritation along the reflex arc in that distribution. Examples of such spasms are blepharospasm, facial spasm, torticollis, etc. A spasm may begin in a single muscle and spread to neighboring muscles. In spasm, the muscles may become hypertrophied from overuse, but this condition never occurs in chorea. (See chapters on Spasm and Tic.) When the chorea is associated with tic or spasm, the diagnosis may be very difficult.

Myoclonias.—Chorea is distinguished from the myoclonias by the fact that in the latter the twitchings occur in paroxysms, and are of lightning-like rapidity (30–120 per minute), clonic in nature, and involve a part of a muscle, a single muscle or a group of muscles, and rarely, if ever, produce movement of the parts involved. They are usually associated with other nervous diseases, especially epilepsy.

Chorea-athetoid Form of Infantile Cerebral Palsy.—Chorea, which has existed from early childhood, may be mistaken for the *chorea-athetoid form of infantile cerebral palsy*. The muscular rigidity, especially of the legs, with other signs of pyramidal tract involvement, the athetoid character of the movements, and the fact that congenital

or early acquired permanent chorea is very rare, will prevent confusion.

Athetoid Movements.—Choreiform movements will not be confused with athetoid movements if it will be remembered that the latter are more commonly seen in spastic, but not completely paralyzed, limbs, and that they are irregular, twisting, "jelly-fish" like in character, involving the distal ends of a limb, and that the face is rarely involved, except in bilateral athetosis.

Associated Movements (Synkinesæ).—Associated movements occur involuntarily in hemiplegics on attempting to execute a voluntary movement with the paralyzed limb. Thus, if the patient attempts to draw up the hemiplegic leg, he involuntarily dorsiflexes the ankle and hyperextends the big toe, or when flexing the fingers, he involuntarily dorsiflexes the wrist. They are usually seen in organic disease of the brain and spinal cord (tabes). Bearing the characteristics of these movements in mind, they cannot be confused with choreiform movements.

Fibrillary Twitchings—Myokymia.—The diagnosis of choreiform movements from fibrillary twitching and from the quivering of the muscles in myokymia will hardly ever present any difficulties (see Tremors).

Tetany.—The spasms in tetany are characterized by being painful and by involving bilaterally the hands and feet, giving rise to characteristic postures ("main de accoucheur," etc.). The presence of Chvostek's, Erb's and Trousseau's signs (see Tetany) will be diagnostic.

Tetanus.—The characteristic tonic spasms of the jaws and face, trunk and limbs, with complete relaxation between the spasms, and the history, will very easily differentiate tetanus from chorea.

Hysteria.—The diagnosis from *hysteria* may be quite difficult, especially when the two conditions coexist. The age and sex of the patient, the absence of a cardiac or rheumatic history, and the presence of stigmata of hysteria, unusual sensory disturbances, a psychic mental make-up, with a history of a sudden onset of the muscular twitchings in isolated parts of the body, their frequently rhythmical character, after the hysterical patient has had an opportunity to observe a patient with genuine chorea, will aid in the differentiation of the two conditions.

Dystonia Musculorum Deformans—Wilson's Disease (Bilateral Degeneration of the Lenticular Nuclei).—These conditions are also characterized by spasmodic muscular twitchings, and the reader is referred to the special chapters on these diseases.

Huntington's Chorea.—Chorea minor is differentiated from Huntington's chorea in that the latter is, as a rule, a disease beginning after the age of thirty, is both hereditary and familial in nature, progressive in its course, with gradual mental deterioration, leading to complete dementia. In the absence of a history of heredity a chronic form of chorea minor will require long observation to make the differentiation between the two conditions positive.

Cerebral and Multiple Sclerosis.—Cerebral and multiple sclerosis, with patches in the cortex, may simulate chorea. They have been

reported as *chorea spastica* and *pseudosclerosis*. The increased reflexes, the rigidity, the chronicity of the disease, and the history, with the age of onset, will make the diagnosis clear.

Complications.—Endocarditis.—Endocarditis is frequently met with in persons who have or have had chorea. Authors differ greatly as regards the frequency of this complication, and its significance. Some consider it a symptom, others an etiological factor in conjunction with rheumatism, and still others a complication. The reasons for the difference of opinion as regards its frequency are: (1) Because the endocarditis may not appear until years after the chorea. (2) In many cases of chorea a murmur, when detected, may be functional and not organic. (3) Because endocarditis may be present without giving rise to any physical signs.

Sturges collected 80 fatal cases and in only 5 of them was the heart normal. Osler states that of 554 cases of chorea, at the Infirmary for Diseases of the Nervous System, 170 presented heart murmurs—149 of these were at the base. Of the 449 cases reported to the Committee on Collective Investigation of the British Medical Association, 113 had cardiac murmurs; how many of these were organic and how many were functional could not be determined.

Stephen MacKenzie examined 33 patients varying from 1 to 35 years after an attack of chorea, and noted signs of undoubted cardiac disease in 60.6 per cent. Osler, out of 140 cases, found the heart normal in 51 cases; in 17 there was some cardiac disturbance which could be considered functional, and in 72 cases or 51 $\frac{3}{7}$ per cent. there were definite signs of organic heart disease. Fagg found heart disease in 17 out of 18 cases of chorea at necropsy; in 5 of these death was not due to the severity of the chorea.

R. S. Eustis,³³ of 60 cases studied in the Children's Heart Clinic at the Massachusetts General Hospital, found 32 or 53.3 per cent. to be choreic on admission, and 10 more gave a history of choreic attacks in the past, making a total of 42 cases or 70 per cent., in which chorea played a part. Abt and Levinson³⁴ found 73 cases of heart disease in 226 cases of chorea, the majority of which were diagnosed as mitral insufficiency, one as myocarditis, two as single aortic insufficiency, and one as double aortic.

In the majority of cases of heart disease complicating chorea, the lesion is one of mitral insufficiency. The murmurs in these patients are due to structural disease of the mitral valve, being produced by the poison of rheumatism, which, as has been pointed out under the etiology, plays such an important rôle in the causation of the disease. The finding of this form of cardiac disease in the cases of chorea, not due to rheumatism or any other infectious agent, is very difficult to explain. A. Ernest Sanson³⁵ considers the signs and symptoms of mitral insufficiency in the non-rheumatic cases different from those which are due to rheumatism. In some of these, he claims, careful examination for many days may fail to elicit any evidence of valvular involvement, but later a soft, slight systolic murmur, localized at the apex, becomes audible; there is

no accentuation of the second pulmonic sound; the ventricles do not become dilated and yet the murmur retaining its original character persists for several years, becoming completely inaudible later in life. Pathological evidence completes the distinction. In these non-rheumatic cases, according to the same author, the left auriculoventricular orifice on its auricular aspect has been found studded and fringed with small papilliform elevations of the endocardium; these are firm to the touch and cannot be detached by rubbing. The endocardium covering these elevations remains perfectly smooth. The vegetations do not begin, as in rheumatic endocarditis, with a change in the epithelium and an attachment to the roughened surface of the fibrous caps, but they are firm outgrowths showing fibrous hyperplasia. This type of endocarditis is not followed by sclerous changes with retraction of the valves, cords and columns, and the endocardium remaining smooth; there is very little interference with the closure of the valve in systole. These changes, says Sansom, "may be the immediate results of a sudden overstrain and rupture of the terminal arterioles distributed to the valve structures." Experimental production of overstrain and fright in animals by Roy and Adami were followed by similar changes in the endocardium.

Pericarditis.—Pericarditis is occasionally met with as a complication of chorea, especially in the rheumatic cases. It is not nearly as common as endocarditis, and presents no unusual features.

Rheumatism.—The rôle which rheumatism plays in the causation of chorea has been discussed under the etiology of the disease. Its association with chorea may manifest itself in three ways: (1) an attack of articular rheumatism will precede by months or years the onset of an attack of chorea, and will not recur before or during the entire course of it; (2) an attack of articular rheumatism will begin simultaneously with the choreiform movements; (3) it will appear during the course of chorea.

Articular rheumatism during chorea is, as a rule, milder in its course than ordinarily. Patients with chorea frequently complain of vague pains in the joints, which are too readily ascribed as due to rheumatism. It is well to bear in mind that while chorea is not a painful disease, and is accompanied with no sensory changes, nevertheless, when the disease is very severe, the constant wriggling and throwing about of the limbs may give rise to pains. Osler asks whether these pains are not analogous to those seen in the limbs and joints during most infectious diseases. The French writers call these pains, for which no local cause can be discovered, *choreic arthropathies*.

Psychoses.—Chorea is frequently complicated with the various psychoses; this is more commonly so in adults than in children. In these cases it is important, both from a diagnostic and prognostic standpoint, to differentiate the existence of a psychosis "per se" from the ordinary psychic states, which are seen in the disease itself. The mental symptoms may be the predominating feature and overshadow the chorea, with the result, as Osler says, that patients have even been committed to the insane asylum.

Feeble-mindedness.—Congenital as well as acquired feeble-mindedness may be combined with chorea.

Epilepsy—Hysteria—Spasms—Tics.—The association of chorea with epilepsy is not as common as with hysteria and the other neuroses, such as spasms and tics.

Cerebral Diplegias and Hemiplegias.—Athetosis and Other Organic Nervous Diseases.—Chorea may also complicate the cerebral diplegias, the hemiplegias and the various forms of athetoses and other organic nervous diseases.

Exophthalmic Goiter.—Choreiform movements may occur in exophthalmic goiter, and the two conditions may be combined.

Anemia.—The association of chorea with the chlorotic type of anemia is so common that anemia may be said to be one of the cardinal symptoms of the disease rather than a complication.

Jaundice.—Jaundice, according to Poynton, is very common in rheumatic children, and it may complicate the course of a severe chorea.

Chorea in Pregnancy.—General Considerations.—The influence which pregnancy exerts on the occurrence of chorea is still unknown. It is commonly regarded as a reflex irritation similar to that which causes the vomiting of pregnancy, but inasmuch as the development of chorea in pregnancy is usually later than that of morning sickness, and rarely ceases immediately after the removal of the products of conception by labor or abortion, the analogy seems hardly tenable. There seems to be no doubt, however, that the fetal movements are the actual exciting cause.

Symptomatology.—The choreic movements may manifest themselves at any time during pregnancy; they are, however, most common in the third or fourth month, and rarely begin in the eighth or ninth month.

The nature of the movements, their onset and involvement are similar to chorea in the non-pregnant. The movements may extend to the uterus. Braxton Hicks³⁶ reports the case of a young woman who had chorea in childhood. During pregnancy the chorea had recurred, and the uterus, which could be distinctly outlined in the abdomen, presented a marked alteration in form, accompanied by evident choreic contractions. Rest in bed, with arsenic, diminished the movements. Labor was uneventful, and she made a good recovery.

Romberg was the first to point out that chorea in pregnant women is more apt to be bilateral, and to involve the tongue.

The psychic manifestations of chorea, in pregnancy, are characterized by greater memory defects and maniacal outbursts than in the non-pregnant. The memory, however, improves with the cessation of the chorea. Maniacal choreics give peculiar outcries, not unlike patients about to be seized with an epileptic convulsion. The irritability, the visual and auditory hallucinations, the dream-like confusion, with the peculiar lack of connection of ideas, do not differ from those seen in the non-pregnant, except that their prognosis is much more unfavorable, because they have a tendency to persist after the chorea has ceased.

After delivery, the choreic movements gradually subside, but they

have been known to continue for 4 or 5 months after labor or abortion, and in a few cases to have remained permanently.

Occurrence.—Recurrences.—Pregnancy always aggravates an existing chorea and predisposes to recurrences. Acute rheumatism is generally the most immediate cause; epilepsy and other nervous disturbances are predisposing causes. Fright, emotion, infection and anemia favor its occurrence. As in the non-pregnant, a recurrent chorea may, during pregnancy, be more severe than a former attack, but in some cases, however, the recurrence may be less severe. Primigravidæ are more susceptible than multigravidæ. The younger the patient, the greater the liability to recurrences. The chorea of pregnancy rarely occurs for the first time in a patient free from rheumatism after the age of twenty-five.

If chorea recurs during several pregnancies, there is no uniformity as to the time of its onset. Lawson Tait cited the case of a patient who was rheumatic, and had chorea at sixteen; she became choreic in the fourth month of her first pregnancy, in the third month of the second, at the beginning of the third, and in the fourth month of the fourth.

There are cases on record in which a first attack of ordinary chorea has been observed to have begun after delivery or after abortion.

Effect of Chorea on Pregnancy and Prognosis.—This depends upon the severity of the disease. In mild cases, if properly treated, the pregnancy may not be interrupted. In severe cases abortion may occur spontaneously. When this occurs, the prognosis is very grave. Barnes³⁷ has collected 56 such cases with a fatal outcome in 1 out of every 3. Most of the patients developed septicemia or pyemia, which was followed by hyperpyrexia, coma and death. The earlier in pregnancy the chorea occurs, the greater the danger for the child. The mortality of chorea in pregnancy, in general, is about 30 per cent.

Diagnosis.—The great liability to hysterical manifestations in pregnancy makes the differential diagnosis in these cases at times quite perplexing. The previous history, the patient's mental make-up, the presence of hysterical sensory manifestations, and the presence of a case of chorea near the patient, with a possibility of "imitation," will make the diagnosis clear.

Treatment.—The treatment of chorea in pregnancy does not differ from the treatment of ordinary chorea. The **salicylates, arsenic, rest, light, diet and hydrotherapeutic measures** are indicated. Trousseau and Gowers have used **strychnin** in physiological doses with success. **Chloral** is a useful drug, particularly in cases with severe insomnia.

Pathology.—The most constant pathological lesions found in the central nervous system are in the region of the frontal lobes and motor cortex. Occasionally the spinal cord may share in the involvement. The nature of the lesions is the same as in the non-pregnant choreas. The same is true of the pathological findings in the other organs.

Obstetrical Indications.—In mild cases no interference is indicated; in severe cases, exhaustion, insomnia, marked mental symptoms or grave physical complications are indications for the artificial termination of pregnancy. It is well to bear in mind that these patients are predis-

posed to hemorrhages, both on account of the generally impoverished condition of the blood and the usual previous administration of large doses of the bromids for the chorea. Owing to the severity of the choreic movements during labor, the second and third stages may be unusually difficult. The patients may actually have to be held down by attendants, or anesthetized, to prevent them from seriously injuring themselves (Kolde³⁴).

The successful cure of pregnancy toxicosis by serotherapy has prompted some obstetricians to use this method in the treatment of chorea in pregnancy. Albrecht³⁵ reports a case in which a patient of his had chorea during her first pregnancy. It had recurred during the second, and treatment with the ordinary methods for 22 days gave no relief; she was then injected with 20 c.c. of serum from a normal pregnant woman, and within 24 hours the chorea that had tormented her for over 3 months subsided permanently. He concludes his report by suggesting that a similar toxic action during the prepuberty stage may be the explanation of chorea in the young; the approach of puberty producing changes in the glands of internal secretions, like the changes inaugurated by gestation.

An interesting case of chorea during pregnancy, bearing on the possible psychogenic origin of the disease, is reported by Flamma.⁴⁰ A woman of 21 developed an attack of chorea in the third month of her first pregnancy. The chorea growing worse, an abortion was induced, after which it gradually subsided. Becoming pregnant a year later, the chorea recurred in the second month. The woman and her husband begged to have an abortion done, as the choreic movements were incessant and severe. Believing that the trouble was principally of nervous origin, a sham abortion under chloroform anesthesia was resorted to, after which the chorea disappeared. The pregnancy was uninterrupted, and she was delivered at full term. In Flamma's review of the literature, he found many cases where no other factor than the pregnancy could be incriminated to account for the chorea. He remarks that pregnancy chorea and chorea in the pregnant are not necessarily the same thing. Sham interruption of the pregnancy proved effectual in another case in his clinic, in which the psychogenous manifestation took the form of uncontrollable vomiting. The two cases with the same origin but different manifestations, cured by the same measure, is believed by Flamma to prove the conception of a psychogenous origin in such cases. He advises before inducing actual abortion, in dubious cases of this kind, to attempt a sham obstetrical intervention.

Chorea Complicating Labor.—The severe type of chorea complicating labor is comparatively a rare condition. The mortality of this complication is very high. In a series of cases recently reviewed, 438 in number, which included all grades of the disease, the mortality in the mothers was 16.5 per cent. The mortality of the newly-born is also very high. Many of them are still-born. It is interesting to note that some of the children which survive may show choreic symptoms shortly after birth.

while others are perfectly normal, and do not show any sign of chorea or other nervous manifestations.

Clinical Types.—**MILD OR SIMPLE FORM.**—In this form, which is the most common, the disease develops slowly and insidiously, rarely suddenly. The children become peevish, restless and irritable; their actions become awkward, they are clumsy and begin to drop things; in school they cannot sit quietly; they write poorly; they pay no attention to their teachers; they begin to bite their finger nails and to pull the buttons off their clothes. They sleep poorly; they have night terrors and enuresis. They shrug their shoulders, twitch their lips, wrinkle their foreheads and wink their eyes. Punishment has no effect; if anything, it aggravates the condition.

As time goes on, they become pale and weak, their appetite poor and capricious, and digestive disturbances become frequent. Headaches and vague pains in the extremities now make their appearance. At this time, the typical choreiform movements become more and more noticeable; at first in one arm, or one leg, or face and arm, or leg and arm, until eventually the musculature of the entire body is involved. These movements may last from three weeks to six months, depending on the severity of the disease. During this time there is usually a period during which the disease is at its height, after which there is a gradual regression of the symptoms. The movements now become less severe and less frequent; later they are observed only during emotional or physical excitement, and finally disappear altogether. The incoördination and weakness disappear before the movements have entirely ceased. In this form, the spasms are slight and speech is almost never seriously affected.

SEVERE FORM.—The disease, as a rule, develops suddenly after fright, trauma or shock. The movements are general, and the involvement extensive at the very beginning of the disease. They are so violent that they interfere with speech and locomotion, thereby incapacitating the patients from dressing and feeding themselves. In this form there may be loss of power in the extremity most involved, giving rise to so-called "paralytic chorea." These cases are usually of longer duration than the milder forms.

MALIGNANT CHOREA OR CHOREA INSANIENS.—This is the gravest and, fortunately, the least common form. The cases belonging to this group may follow the simple or severe form, or they may be the terminal stages of these forms, or they may begin as such. In the latter cases, it is seen most frequently as we approach adult life. This type is characterized by a predominance of the mental over the hyperkinetic symptoms. The patients become maniacal, delirious and confused; they have delusions and hallucinations; they cannot sleep, the temperature rises to 104° F. (40° C.) and sometimes as high as 106° F. (41.1° C.), and death is the usual termination. The psychic symptoms in this type are so marked that they are occasionally mistaken as symptoms of an infectious or toxic psychosis. The simple delusional states which one meets at times in the other forms of the disease are insignificant in

comparison with those seen in this grave form, and can hardly ever be confused with them.

Treatment.—GENERAL MEASURES.—Because the disease is one of long duration, it is important to keep the patient under the best general hygienic conditions possible.

HYGIENIC MEASURES.—Localized infectious foci should be removed, diseased tonsils excised, discharging ears treated, decayed teeth extracted, and general oral hygienic measures resorted to. The food should be abundant, light and nourishing. In cases complicated with rheumatism, red meats should be given sparingly. Plenty of water and fresh or stewed fruit are indicated. In cases with intestinal indigestion, the daily irrigation of the intestinal tract is advisable. Lactic acid cultures have been found to be of value in such cases. Tea, coffee and sweets should be used moderately.

Rest.—Absolute rest is essential, and in severe cases isolation with a tactful nurse indicated. Excessive mental or physical exercise is to be forbidden. Ambulatory cases are not to be allowed to attend theaters or moving picture exhibitions.

MEDICINAL TREATMENT.—The three most important drugs in the treatment of chorea are arsenic, antipyrin and the salicylic acid group. **Arsenic** has been employed with success as far back as 1870 by Alexander, and recently by Pawlow, Bokay, Marie and others in the form of salvarsan. In the treatment with arsenic it is important to bear in mind the untoward effects of the drug. Arsenical neuritis, gastritis or nephritis may result from the injudicious use of the drug and may then be as serious as the chorea itself. The writer believes the administration of arsenic in the following way will minimize its possible untoward effects: For a child eight years old begin with 4-5 drops of Fowler's solution, well diluted, three times a day after each meal, to be increased daily by one drop till 12-15 drops have been taken three times a day. Seguin⁴¹ regards 18-27 drops three times a day a normal dose. The parents are instructed to watch for nausea, vomiting, pain in the stomach, puffy eyelids and severe neuralgic pains in the extremities on the days when the larger doses are given. If such symptoms do occur, say on the sixth day, it is established that this particular patient's maximum dose is ten drops three times a day. After this, the patient receives no arsenic for three or four days; this gives him an opportunity to get rid of the untoward effects of the arsenic already ingested, after which the original dose of four drops, three times a day, is resumed and gradually increased till he obtains ten drops t.i.d.—the maximum dose, which he can apparently tolerate.

Eulenberg and Hammond have used arsenic hypodermatically. Bokay⁴² administered 0.2 gram (3 grains) salvarsan subcutaneously to a child 8 years old; improvement began on the second day; it was striking on the fifth, and after four weeks recovery was complete. Another child with a less severe chorea under treatment with Fowler's solution made a much slower recovery. Two cases with recovery in 15 days after the injection of 0.2 gram (3 grains) are reported by

Hainiss,⁴⁸ and Benno Hahn⁴⁴ reports three cases with intravenous injection of 0.08-0.3 gram (1.3-5 grains) and recovery in from 8 to 28 days. Each of Hahn's patients received three injections, three to eighteen days apart.

We have used sodium cacodylate intramuscularly and neosalvarsan intravenously, with no better results than with Fowler's solution by mouth. Arsenic may also be used by mouth in the form of arsenious acid (grains 1/130-1/60); its combination with iron in cases in which anemia is a marked feature, and, with digitalis in cardiac cases, is advisable.

Antipyrin has always been a favorite remedy for chorea with foreign clinicians. It is best administered in doses of 5-8 grains (0.324-0.52 gram), three times a day. Comby always uses antipyrin in very large doses, 90-140 grains (5.85-9.10 grams) a day, and if the disease does not respond to these doses, he employs arsenic.

Salicylates are indicated when chorea is complicated with manifestations of rheumatism; they are best administered with alkalis to prevent gastric irritation. Langmead uses salicylates only when the disease is accompanied with very high temperatures; he does not believe that this drug has any effect on the severity or duration of the chorea. Pearce Bailey⁴⁵ advises the use of **aspirin**, whether the cases are complicated with rheumatism or not, but he insists on the importance of absolute rest and isolation, with the application of cold packs and occasionally lumbar puncture, when the fluid is under high pressure. He advises, after the movements have ceased, a three or four weeks' stay in the hospital for the purpose of reestablishing the tone of the nervous system, and thereby diminishing the tendency to a relapse. He very rarely had occasion to use salvarsan.

For the motor restlessness, **chloral**, **bromids**, **belladonna**, **hyoscyamus**, **hyoscyamin**, **hyoscin**, **apomorphin**, **gelsemium**, **cannabis indica**, and even small doses of **morphin** may have to be administered. **Sulphonal**, **trional**, **chloreton** and **paraldehyd** may be employed when the insomnia is wearing out the patient. In this connection it will be interesting to note an experience which Gairdner⁴⁶ had with chloral in the case of an eight-year-old girl with chorea, who took by mistake sixty instead of twenty grains of chloral to induce sleep. She recovered from the poisonous effects of the overdose of the drug and was permanently cured of her chorea.

Sachs recommends tincture of **cimicifuga** in doses of 15-30 drops, three times a day, especially when arsenic cannot be tolerated. Mayer injects intravenously small doses of **phenosol** in 1 per cent. strength with good results, he claims. Grober never saw any good from salicylates or any other drugs; he depends mainly on early and thorough purging, complete rest and prolonged warm baths (93.2° to 98.6° F. [34° to 37° C.]), three daily; he is opposed to the pack because it tends to irritate the patient. In cases with cardiac complications, he considers the application of an ice bag to the heart a dangerous procedure, and em-

plays moist heat to the precordial region; he urges unusual care during convalescence to prevent neurasthenia.

INTRASPINAL AND SUBCUTANEOUS INJECTIONS OF MAGNESIUM SULPHATE.—Meltzer and Auer, of New York, have shown that magnesium sulphate in intravenous, subcutaneous and intraspinal injections and local applications to nerve trunks inhibits the excitability and conductivity of nerve tissue. They then suggested the subarachnoid injection of this drug as a substitute for cocain in producing regional anesthesia. The first therapeutic use made of this fact was in the treatment of tetanus. Marinesco then demonstrated the good effects of magnesium sulphate when injected into the subarachnoid space in sciatica and in the gastric crises of tabes, and later he applied this method of treatment to Sydenham's chorea. In 1908, he reported 4 cases of this disease, treated successfully by this method. Good results were later reported by Baduel, Rocaz, Calcattera, Caronia and others. Augusto Natali⁴⁷ reports the successful treatment of eight severe cases of chorea by the so-called Marinesco method. His technic was as follows: He performed lumbar puncture in the usual manner, evacuated a certain amount of cerebrospinal fluid, and injected a solution of magnesium sulphate; the strength of the latter varying from 7 to 25 per cent. and the amount injected was 1 c.c. for every 25 pounds of body weight. All the cases had previously been subjected to the usual methods of treatment, without any signs of improvement. After treatment with magnesium sulphate intraspinally, in 6 of the cases, the results were remarkable; in 2, one of which was a chronic case, there was also a marked improvement, but it was very slow. Natali points out the advantage of following the treatment by means of a brief course of arsenical medication; he does not believe that the magnesium sulphate acts as a specific, but is merely a symptomatic remedy, diminishing the excitability of the nerve centers.

Excellent results have been reported by R. Pastore⁴⁸ in 4 cases of chorea in which magnesium sulphate was given intraspinally in small repeated doses. She gave 5 to 7 injections of 0.1-0.2 gram (1.5-3.0 grains) of magnesium sulphate in a 25 per cent. solution to a total of 0.4-0.5 gram (6.0-7.7 grains); the intervals were from one to two days at first and three to seven days later. There was no reaction; one child improved after the third injection; another child after the sixth and another after the seventh. In one little girl of eleven, the third injection was followed by severe headaches, superficial breathing, sluggish pupils and a return of the choreic movements, but, upon two more injections given after this, pronounced improvement set in.

Heiman,⁴⁹ of New York, treated five successive cases of chorea with repeated *subcutaneous* injections of magnesium sulphate. In every case a 25 per cent. sterile solution was used; the dose ranging from 0.01 gram (1/6 grain) per kilogram of body weight, i.e., 0.04 c.c. of the 25 per cent. solution at the beginning of the treatment, with a daily increase to 0.2 gram (3 grains) per kilogram of body weight, i.e., 0.8 c.c. of the 25 per cent. solution at the last injection. The actual amounts of the solu-

tions used daily were from 3 to 30 c.c.; they were given three times daily for from 10 to 15 days, with the ordinary Record syringe in the back, loins and buttocks. In only one of the five cases was there a noticeable improvement after the series of injections, and in this one case the movements subsided gradually. Not only did the remaining four cases show no improvement, but they all had severe inflammatory reactions after the injections, and in one of them a marked albuminuria set in.

In marked contrast with Heiman's experience was that of Schroeder,⁵⁰ who used a 20 per cent. solution of magnesium sulphate subcutaneously in nine children, and two adults with severe chorea, with such good results that he recommended this method to be used in all hospitals.

SERUM TREATMENT OF CHOREA.—Goodman, of New York, investigated the etiology of chorea and noted pathologically the involvement of the central nervous system; this suggested to him that the disease was infectious in nature and that the older methods of treatment had failed because the remedies had not reached the seat of the disease, so he began to use the **patient's own serum** intraspinally. His technic is as follows:

- (1) Exclude syphilis and tuberculosis.
- (2) Rest in bed for several days without treatment in order to eliminate all previously used drugs from the system.
- (3) Blood, to the amount of 40-50 c.c., is drawn from a vein and centrifuged; this amount of blood will furnish about 15-20 c.c. of serum. Keep on ice till ready for use.
- (4) Lumbar puncture is performed and 15-20 c.c. of spinal fluid is withdrawn.
- (5) The serum is warmed to body temperature and from 10 to 18 c.c. is injected into the spinal canal; this is done slowly, the average time consumed being from ten to fifteen minutes.

- (6) The patient remains flat on his back for an hour or two.

Of 30 cases treated by Goodman, according to his original report, 14 received one injection, 8 two, 5 three, and 3 received four injections. Of those receiving 1 injection, 12 were cured and 2 markedly improved; of those receiving 2 injections, 5 were cured and 3 markedly improved; of those receiving 3 injections, 2 were cured, 1 markedly improved, 1 slightly improved and 1 unimproved. Of those receiving 4 injections, 1 was cured, 1 markedly improved and 1 unimproved.

Tarr⁵¹ treated 14 patients with Goodman's "auto-serum method"; 11 of the 14 cases were cured, 2 improved and 1 unimproved; total number of injections, 23. The greatest number given to a single patient was 3; the amount of serum, per dose, ranging from 7 c.c. to 18 c.c. The youngest child of the series received 16 c.c.; the oldest, which was 16 years old, the same dosage. The former was well in 48 hours, while the latter was improved in 4 days and was completely cured in about two months. Tarr gives no solid food for the first 12 to 24 hours, thus avoiding nausea and vomiting, and excludes all visitors for 48 hours, leaving the patient in a quiet room, alone as much as possible. Anesthesia may be used in severe cases when necessary. Tarr con-

cludes that whatever merit there may be in the treatment, it certainly can do no harm.

H. K. Faber⁵² employed the so-called Goodman method in 3 cases, and almost immediate cessation of the choreic movements followed the first injection, with no recurrence of symptoms in a little over a month.

Porter,⁵³ of San Francisco, reported before the American Pediatric Society his experience in the treatment of chorea by means of **intrathecal injections of horse serum**. Seven cases were injected intrathecally in the ordinary way, and one by the method which Mehrtens of the Stanford University employs in the treatment of cerebrospinal syphilis. (Mehrtens injects arsphenamin from six to twelve hours after an initial dose of horse serum of 0.5 c.c. is given in order to find out whether the patient is hypersensitive to horse serum.) Five of the patients received a second injection on the fourth, fifth and sixth days following the initial dose; these injections were followed by strikingly rapid improvement in most cases, but no case ceased to twitch absolutely within a week; in fact, only in the mild cases was the twitching absent in two weeks. Porter admits that, on the whole, his results do not encourage the hope that this is any advance on other methods of treatment, and he would only advise the treatment as of value in controlling very severe cases, but not in moderately severe cases.

Oscar Schloss, of New York, employed the same method in 12 cases, but he could find no difference in the results from twelve control patients who did not get the serum. La Fetra is not enthusiastic over either this (horse serum) or the Goodman method.

There is no doubt that the carrying out of these methods presents considerable difficulties and requires great technical skill. The evidence of other competent observers in addition to that cited is such that, although the writer's personal experience in this field has not been very extensive, he believes that their therapeutic value must still be held *sub judice*.

Haneborg⁵⁴ suggests that, inasmuch as chorea is most prevalent in children between the ages of two and sixteen, i.e., the period of thymus gland activity, the thymus must contain some secretion or secretions which have a tranquillizing influence on the nervous system, and that chorea must be due to the lack of these secretions. With this view in mind, he has employed **thymus extract** in the treatment of chorea, he claims, with satisfactory results.

Timme⁵⁵ reports a case of severe infectious chorea in which nothing was done for the patient, except **absolute rest**; after four days **lumbar puncture** was done, and 25 c.c. of fluid, which was under high pressure, removed. At the end of 24 hours, the patient, although still suffering from the tonsillar infection, became normal, as far as the chorea was concerned. There was no return of the chorea. Another child with rheumatism, tonsillitis and chorea was subjected to the same treatment, and in 24 hours after the withdrawal of the fluid the chorea partially subsided, although the child still had rheumatism and tonsillitis. Thirty c.c. more fluid was withdrawn, and the choreic movements

ceased absolutely and permanently. Strauss, of New York, in attempting to follow the same procedure as a therapeutic measure was unable to obtain uniform results. The fluid was not under high pressure. Morse and Floyd found no noticeable effect at any time on the choreic symptoms after lumbar puncture. Indiscriminate lumbar puncture in patients with chorea is dangerous; we have seen several cases in which the chorea became more violent after the lumbar puncture, the patients becoming more nervous than they had been before.

Various **injections with vaccines** (stock and autogenous) and sera are being used by different clinicians. Inasmuch as it was pointed out under the etiology, the bacterial origin of the disease as well as its pathogenesis in general have not been definitely determined, there is at present no scientific basis upon which the writer can recommend the use of these procedures.

MECHANOTHERAPY—KINESITHERAPY.—In the milder and in the chronic forms of chorea, **mechanical treatment** by means of **suggestion** and **passive movements** at first, and later by means of **exercise**, has been worked out and systematized by Guthrie. The movements employed are very simple; at first slowly and steadily, like in Frankel's method in the treatment of tabes. Grossman⁵⁸ claims to have treated successfully 50 cases of chorea by breathing, relaxation and reëducation exercises. He also begins with the simplest possible movements, and as the patient is beginning to show signs of improvement, he orders more elaborate exercises; by this method he says the duration of the disease is shortened, and residual chorea obviated and complications lessened. We have had no experience with this method.

HYDROTHERAPY—ELECTROTHERAPY.—**Mild hydrotherapy** in the form of warm packs followed by cold spongings or warm tub baths lasting for one or two hours are sometimes very beneficial. Some physicians have found **electricity**, in the form of galvanism to the back, head and pressure points (if any can be found), useful. In our experience, unless the patient is an adult and the disease of long standing, electrical treatment of any kind has a tendency to aggravate the condition.

TREATMENT DURING CONVALESCENCE.—In convalescence, gymnastic exercises, a change of scenery and a sojourn at the seaside or in the mountains will hasten to establish a cure.

INSTRUCTIONS TO PARENTS.—It is wise to impress upon parents the importance of **clothing** their children **properly**; not to consider lightly frequent attacks of *sore throat*; to pay greater attention to so-called *growing pains*; to remember that *night terrors*, *nervousness*, *clumsiness*, *restlessness*, and *incontinence of urine* may be the first symptoms of an impending attack of St. Vitus dance, and lastly, that *rheumatism* and *chorea* both have a great tendency to recur.

Prognosis.—The ordinary outcome is a complete cure, but in rare cases a permanent loss of muscle power or a weakness of the mental faculties may remain. When these occur they are probably due to organic changes in the nerve centers, which have been brought about by the chorea. The mortality in Pineles' experience is 2.4 per cent.; in

the cases of the British Chorea Committee the mortality was approximately 2 per cent.

The prognosis is much more favorable in children than in adults. It is more serious if the chorea follows immediately, or soon after, any of the infectious diseases. In the very severe cases, the cases of chorea gravis (chorea acutissima), the prognosis is very bad: most of the patients die within a week or two. A guarded prognosis must be given in cases complicated with severe mental symptoms, or definite, organic cardiac lesions. The same is true of cases in which hyperpyrexia is a prominent symptom. The appearance of an exanthem, resembling scarlet fever, during the course of the disease, is an unfavorable sign.

The prognosis is somewhat better in a recurrence than in a first attack.

When death occurs, it is usually due to cardiac failure or to exhaustion from insufficient nourishment and lack of sleep, or to injuries which the patients might have sustained during their violent and incessant jactitations.

Pathology and Pathogenesis.—The most constant lesions found on autopsy are endocarditis, the mitral valve being the one most commonly involved, fatty myocardium with Aschoff bodies (submiliary nodules in the wall of the left ventricle) and ulcerative endocarditis with metastatic emboli in other organs.

The changes found in the brain, spinal cord and nerves are as varied and as numerous as those who describe them.

Kopezynski found no changes in the brain. Starr, who believes that the disease is purely functional, ascribes the postmortem findings in the central nervous system as secondary in nature. Gowers and others also believe these changes to be secondary in nature. Bechterew thinks that the disease is infectious, but whatever the infectious agent may be, it does not produce gross lesions in the nervous system.

Hughlings Jackson thought that the almost constant involvement of the facial muscles proved the cerebral origin of chorea, and so he elaborated the so-called "embolic theory" of chorea. The association of endocarditis with chorea was pointed out by him and his followers as evidence in favor of his theory, but the failure of other competent observers to find emboli in fatal cases, and the fact that there are as many cases of chorea without endocarditis as with endocarditis seem to be sound arguments against the embolic theory; as a matter of fact, terminal emboli have been found with more frequency in the central artery of the retina than in the cerebral capillaries.

Choreiform movements have been produced experimentally by Rosenthal and by Angel Money⁵⁷ by injecting fluid into the carotids of animals, as a result of which, capillary emboli were found in both the brain and cord. Stimulation of the motor cortex with chemical poisons, especially creatinin, also was followed by chorea.

Dickinson⁵⁸ found, as the most common pathological condition, a hyperemia of the brain and cord with hemorrhages in that part of the brain which is supplied by the middle cerebral artery.

Horatio C. Wood believes, as a result of his experiments in animals, that the pathology of chorea is a change in the nutrition of the ganglionic structures of the entire cerebrospinal axis, and that this change in nutrition may in some cases fail to develop structural changes sufficiently great to be detected by the microscope, while in other cases it may go on until it produces pronounced structural lesions.

Meynert⁵⁹ and Elischer⁶⁰ found hyaline degeneration in the nerve cells of the central ganglia. Flechsig also found hyaline changes in the anterior part of the lenticular nucleus. Elischer, Jakowenko and others found, concentrically situated, strongly refractive granules, the so-called "chorea corpuscles" in the vessel walls of the lenticular nucleus, but Wallenberg and others do not consider these characteristic of chorea, because they have also been demonstrated in diseases of the brain other than chorea.

Alzheimer⁶¹ reports changes in the corpus striatum and the subthalamic region in two cases of chorea, associated with sepsis; and in two cases of "rheumatic" chorea there were small foci of proliferated glia cells with rod-shaped cells, particularly near the vessels, and in the septic cases, heaps of cocci occluded the vessels. In the "rheumatic" cases there were similar foci of cells but no microorganisms could be demonstrated. He thinks, therefore, that septic and rheumatic chorea both depend on embolic foci, which are most frequently situated in the corpus striatum and subthalamic region.

Lucien Libert⁶² reports a case of chorea in a patient with a family history of choreiform movements, and a personal history of alcoholism, in which on necropsy two small tumors were found in the dura of the frontal region. He ascribes the choreic symptoms to the tumors in this localization. There are other cases of chorea in the literature, without a history of alcoholism or heredity, in which similar lesions were present, but their clinical history is such as to cast a good deal of doubt as to whether they were cases of Sydenham's chorea.

B. Conos⁶³ cites the case of a woman of 77 with a history of epileptic seizures who, on admission to the hospital, exhibited marked choreic movements confined exclusively to the left side of the body, involving at first the extremities and later the head and neck; the reflexes were lively but normal, and there was no Babinski and no clonus. The urine contained albumin but no sugar. She died of a carbuncle. The microscopic findings in the brain were: (1) general arteriosclerosis; (2) thickened meninges in the left Rolandic area; (3) a focus of softening 15 sq. mm. in front of the left parieto-occipital fissure; (4) a slight atrophy of both frontal lobes; (5) the entire left lenticular nucleus was diseased and in the center of the lesion a small cavity, around which the brain substance was soft and of a yellowish color; (6) the right hemisphere showed nothing but a small focus of disintegrated tissue, of the size of a pinhead, on the internal surface of the globus pallidus, a lesion to which Conos attaches no significance. The chorea on the same side as the lesion he explains as due to a congenital absence of the pyramidal decussation, such as had been seen by Bidon

(published by Pitres), by Zenner (cited by Oppenheim) and by Dupré. From the study of this case, he concludes that lesions of the lenticular nucleus give rise to choreiform movements. Inasmuch as there was no anesthesia and no astereognosis, he is positive that the lesion was confined absolutely to the lenticular nucleus. He quotes Henri Claude as supporting his theory, and also cites several autopsies in which the findings sustained it.

J. R. Hunt⁶⁴ believes that lesions involving the small ganglion cells of the caudate nucleus and putamen (neostriatum) give rise to chorea, because the function of this system of cells is inhibitory and coördinating, and loss of these functions in these cells by disease gives rise to the wild and irregular movements.

Steiner, Hutchinson, Clarke and others are of the opinion that lesions of the spinal cord are responsible for the choreic movements. They base their opinion on the anemia and proliferation of connective tissue which they found in the upper part of the cord.

Changes in the axis cylinders of the peripheral nerves have been found by Elischer, and changes in the muscles in paralytic chorea by Rindfleisch. Gowers, Bonnhoeffer and Foerster think the cerebellum, or the tract of the superior peduncles, is the starting point of the disease. Oppenheim states, "There can be no doubt that chorea is a brain disease, but we cannot say with certainty whether the lesions are localized in the central ganglia, the cerebral cortex, the cerebellum or in all of these."

Damaye⁶⁵ believes chorea to be an organic brain disease, due to encephalitis. Comby⁶⁶ thinks the disease is a neurosis of the same character as epilepsy, and that both of these are most commonly relics of an acute encephalitis, i.e., they are both acquired, accidental maladies. Choreia he believes to be an expression of a mild, acute, curable encephalitis of infectious origin, and that almost any infectious disease may provoke it. Gareiso⁶⁷ reports signs of encephalitis in 49 out of 50 cases of Sydenham's chorea, tested for the Babinski and other signs of organic disease; the reflex responses varied from day to day, and as the chorea subsided, the signs of the superficial transient encephalitis subsided with it.

Runge⁶⁸ points out that inasmuch as most of the psychoses occurring in chorea are similar to the types seen in the exhaustion and infectious psychoses, certain weight is lent to the theory of the infectious origin of the disease. Schiotz,⁶⁹ from a study of 211 cases, concludes the condition to be a neurosis which develops in those predisposed to it between the ages of eight and twelve, and that the inciting cause may be either toxic, infectious or emotional stress.

Summary of Etiology, Pathology and Pathogenesis.—The evidence presented in the previous pages, as to the etiology and pathology in chorea, shows that up to the present time there is no definite scientific basis for the pathogenesis of the disease..

The apparently contradictory etiological and pathological findings are probably due to the fact that the different writers, in describing

chorea, confuse *Sydenham's* or *infectious chorea* with the *degenerative choreas*, or choreiform movements due to organic disease of the brain, or as a neurosis reflected from some remote pathological condition.

Considering Sydenham's chorea in the sense that it is described in this chapter, i.e., a distinct clinical entity, one is justified in making the following statements as regards the probable pathogenesis of the affection:

(1) That, in spite of the studies of Macalister (*see* under Etiology), from which it would appear that the poisons of *rheumatism* and *chorea* are not similar, there is more than abundant clinical evidence that rheumatism, chorea, and some forms of endocarditis are closely related to one another.

(2) That the gradual or sudden onset of the disease, its slow progressive course, with occasional fever, its tendency to recurrences and to cardiac and arthritic complications, as well as the similarity of the psychic symptoms, when such exist, to those of the infectious or toxic psychoses, point to a probable toxic or infectious origin.

(3) That whatever this agent may be, whether infectious or toxic, or both, it has hitherto not been demonstrated, but bacteriological investigations seem to indicate that it is most probably a bacterium whose cultural characteristics are still to be determined.

(4) That the toxin or toxins elaborated by this bacterium has a predilection in those predisposed, for nerve, joint and cardiac tissue, just like the toxic agent in what is known as Wilson's disease, has a predilection for the lenticular nucleus, hepatic cells and the cornea.

(5) That, while there is no doubt that chorea is a brain disease, the exact localization for the motor phenomena has not as yet been determined, but that it is most probably due to involvement of the motor cortex, the basal ganglia, the cerebellum, or all of these.

Historical Summary.—The names of St. Vitus's dance, the dance of St. John, chorea minor, chorea major, and chorea germanorum, have been used with varied meanings in regard to their significance. It appears that the Phrygian bacchantes, in their wild worship, were affected with violent automatic movements, accompanied by more or less disturbance of consciousness, and it is certain that the sect of the Suffi, in Persia, shortly after the origin of Mohammedanism, were accustomed in their sacred ceremonies to pass into a condition of wild excitement with dancing, muscular spasms and general convulsions. About the year 1000, a sect of the Suffi found numerous followers and imitators throughout Asia Minor, in Persia, Egypt and Greece. In Christian countries the so-called dance of St. John was already, at the time of the Crusades, an observed custom. It was not until the outbreak, in 1418, of a fresh epidemic of religious excitement in Strasburg that the term "dance of St. Veit" began to be freely applied to these religious disorders, because during this outbreak the chief magistrate of Strasburg ordered those afflicted with dancing mania to repair to the chapel of St. Vitus in Zabern, a village in Alsace, near Strasburg. The name St. Vitus appears to have had its origin from St. Veit, a boy

who, born in Sicily, suffered martyrdom in the year 303 during the persecution of Diocletian, and whose body was carried from place to place, until finally it was buried in the cloister of Korvey.

Paracelsus called these epidemics "*chorea sancti viti*" and "*chorea lasciva*." The name, St. Vitus dance, is the only point of affinity between the old religious dancing manias and the "idiopathic" chorea of the present day. "Idiopathic" chorea was placed on a firm, scientific footing by the classical description of Sydenham. Many German writers speak of the affection in childhood as *chorea minor*, while the term *chorea major* or *chorea germanorum* is used to express affections resembling those of the hysterical epidemics of the Middle Ages. Sometimes the term "*chorea major*" or "*chorea germanorum*" is used to designate what we ordinarily understand to-day by "*hysteria major*."

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HUNTINGTON'S CHOREA

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Synonyms.—Hereditary chorea, Chronic progressive chorea, Choreic dementia, Huntington's disease, Degenerative chorea.

Definition.—A chronic progressive hereditary disease, appearing rarely before the end of the third decade of life, and characterized by irregular choreic movements, speech defects and gradual dementia.

Etiology.—**Sex.**—The prevailing opinion seems to be that both sexes are about equally affected, although Huntington thinks that the disease is more prevalent in males. Wollenberg's statistics bear out Huntington's claims.

AGE.—Cases of Huntington's chorea have been reported in childhood. Stevens reported a case which began in infancy, but it is questionable whether it was a case of true Huntington's chorea. Jolly saw a case in which chorea and epilepsy developed at the age of 9 years. Osler saw a case at the Johns Hopkins Hospital at the age of 18. Hoffman, Peretti and others have reported cases which began in the second decennial period of life. Mackey had 2 or 3 cases below 30. These and similar cases are exceptions; the disease is preëminently one of adult life beginning most frequently in the middle period of life, or early part of the second half between 30 and 40. It rarely commences in old age. It seems that the age of onset bears some relation to the course of the disease; there are certain strains of families in which the age of onset is earlier than in others. According to Heilbronner, when the disease descends through several generations, it has a tendency to develop later and later in life.

HEREDITARY INFLUENCES.—The disease is transmitted from one generation to another; one generation may be passed over, or its members may suffer from insanity, epilepsy or hysteria instead of chorea. From the statistics at hand, it would appear that if children of choreic ancestors get through life without any manifestations of the disease, the thread is broken and the grandchildren and great-grandchildren may be assured that they will be free from the disease.

Muncie, a field worker for the Eugenics Record Office, was set to collect statistics on the disease (originally started by Jelliffe). She was able to construct 4 great pedigree charts containing 441 female and 521 male choreics—962 cases in all, and in addition 10 cases of Sydenham's chorea. The entire number of individuals studied was 4,370. The relatives, even those who did not have chronic chorea, had other nervous diseases. Thus, epilepsy occurred 39 times; infantile convulsions, 19 times; meningitis and encephalitis, 51 times; hydrocephalus, 41 times; feeble-mindedness, 72 times; Sydenham's chorea, 11 times; and ties, 9

times, mostly in one small family. These 962 cases originated from 6 or 7 ancestors who settled in eastern Long Island, south-central and southwestern Connecticut, and eastern Massachusetts; from these localities the disease has spread along the lines of immigration as far as the Pacific coast—one case having been found as far West as Los Angeles. The disease has been handed down in these families, without a break, through four generations; the heredity having been recognized, there have been a few individuals who voluntarily abstained from marriage, but this abstention was not very marked. Among these families people of high mental accomplishment were by no means rare; they included legislators, professors, ministers, authors, one judge and one eminent surgeon. Some, however, broke down later in life, many of them showing lack of responsibility, immorality, and a tendency to alcoholism.

Lewis¹ reports a case of Huntington's chorea in a man of 50, living in Allegany County, in the State of New York, the members of whose family have always recognized that they were liable to diseases of the nervous system. An older brother of the patient had a similar condition which was never diagnosed; he died insane. One younger brother has a similar condition, but not so marked; one sister is neurotic, but has neither motor nor mental symptoms. The mother died at 60 of pneumonia, after having suffered from similar symptoms, and it is believed, although not positively known, that the patient's maternal grandfather had the same trouble. One collateral female cousin has the disease in a very severe form; other cousins could not be traced.

De Castro reports two typical cases of the disease in two individuals with no history of heredity in either family, and they both have perfectly normal children. The writer has now under observation 2 cases, in neither of which any hereditary relationship as to chorea can be ascertained, although both give a bad family history of neuropathy.

The disease bears no etiological relationship to Sydenham's chorea.

Symptomatology.—MODE OF ONSET.—Huntington's chorea has, as a rule, a gradual onset. *Premonitory symptoms* in the nature of mild muscular twitchings, clumsiness in movements requiring coördination, and various manifestations of the neuroses, and abnormal mental states may occur for years before the typical, irregular, coarse jerkings make their appearance. *Emotions* have a very important influence upon the development of the disease. Traumatism and pregnancy seem to be common exciting causes. Horstman² reports a case in a man who developed the disease suddenly after he had fallen off a horse and sustained an injury to his head; but two years before the injury, without having shown any choreiform movements, his mental condition became very poor. He committed suicide, and on postmortem, typical lesions of Huntington's chorea were found.

OBJECTIVE SYMPTOMS.—The most striking part of the clinical picture, and the one without which the disease cannot be recognized, are the peculiar, sudden, purposeless, jerky movements of the head, trunk and extremities. The movements are those of extension, flexion and grasping; there is a worm-like spreading of the fingers, somewhat similar

to athetoid movements, but much quicker in rhythm. The patients "pucker up" their lips as for sucking or whistling; they wrinkle their foreheads, dilate the nostrils, open and close their eyes, and perform "rolling" movements with their tongues. This constant varying play of involuntary movements gives rise to a series of peculiar gesticulations and grimacing, and the peculiar speech. As a rule, the spasms begin in one of the lower extremities, and extend to the upper extremities and to the face; they may remain confined to one extremity for a considerable period. According to Hamilton, the heart, the muscles of respiration, the bladder, the stomach and scrotum may participate in the movements. Psychological excitement intensifies the movements; during rest they become less marked, and usually, though not always, cease entirely during sleep.

The patient can for a short period inhibit these choreic movements by voluntary effort or movement; he thus is able to eat, write, grasp an object, thread a needle, etc. The author, on one occasion, saw a patient with the most violent spasms shave himself with an ordinary razor. This voluntary inhibition of the choreic limbs is often at the expense of increased activity in the other muscles, and the voluntary movements are carried out, not regularly, but only at certain times, the chorea reappearing in the intervals.

Muscle power is, as a rule, undisturbed; rarely is it diminished and occasionally there may be a hemiparesis. The myotatic irritability is increased; the striking of an individual muscle results in the raising of a welt, which it takes a few seconds to disappear. In some, but not in all cases, the continuous movements may be followed by considerable fatigue and exhaustion.

As the disease progresses, the limbs become somewhat rigid, and although the patients are still able to walk, they develop a peculiar staggering gait. The upper part of the body seems to advance ahead of the pelvis and legs, the trunk assumes a rocking movement, the arms swinging from side to side. The legs seem to skip steps, and locomotion in general is carried out very quickly and irregularly; the gait often resembles that of a drunken man—a cerebellar gait. At this time, a slight tremulousness of the hands is superadded to the choreiform movements; Romberg's symptom may often be elicited.

The face becomes expressionless, and assumes the most bizarre contortions. Slight ptosis of the lids may be noticed.

The writing, earlier in the disease, is tremulous and irregular, but later it cannot be carried out at all.

The reflexes are usually slightly exaggerated; very rarely diminished. The pupils react to light and accommodation. There are no characteristic changes in the fundi. The blood may show a slight anemia. The cerebrospinal fluid and urine are normal.

Sensation is not disturbed, but on account of the psychic dullness there is a tendency to general hypesthesia. Some patients complain of intense headaches. There is no sphincteric involvement.

Some patients have a ravenous appetite; they suffer a good deal from

insomnia. According to most observers, their sexual desire may be increased (Hoag).

MENTAL SYMPTOMS.—There is a progressive weakening of the processes of perception, combination, memory, attention and judgment. The patients are excitable, aggressive, and dull emotionally. Absent-mindedness is a characteristic and early symptom of the disease; this absentmindedness is often wrongly interpreted as dementia, which does not really exist. They often have delusions of persecution and phobias; they seldom have euphoria or delusions of grandeur. They are more commonly depressed, and the tendency to commit suicide is marked. Some patients are very loquacious and may have attacks of transitory mania; they are all very restless. As the disease advances they become disoriented as to time, place and person; all mental faculties are in abeyance, and the restlessness is replaced by apathy. The mental weakness may come before or simultaneously with the motor symptoms, but generally appears some years after. There are some rare cases in which the intelligence is not noticeably impaired.

Diagnosis.—The following traits are characteristic of the disease: (1) direct heredity; (2) persistent tremors of head, trunk and limbs; (3) onset of the tremors in middle or later life; (4) the progressive nature of the tremors; (5) progressive mental deterioration. These five diagnostic criteria are often found together, but in a good many individuals some of them may be absent. Several members of a family, or two or more generations, will show a specific symptom-complex for that family. The choreic movements rarely skip a generation, and are, therefore, considered a predominant trait. In some families a psychic disorder of a hyperkinetic type, i.e., mania, may be a predominating trait.

There are thus various biotypes of the disease: there is a biotype without tremors but with mental weakness; another with tremors and no mental weakness; another in which the onset of the motor symptoms is early in life; another in which there is a considerable interval of time between the development of the mental symptoms and the choreiform movements. Different strains of families have different symptom-complexes, so that the age of onset, the degree of motor restlessness and the extent of mental degeneration will show family differences, and will enable one to recognize various biotypes of the disease.

DIFFERENTIAL DIAGNOSIS.—In general, it may be said that if the hereditary nature of the disease cannot be definitely established, the symptoms alone are not sufficient to distinguish the disease from simple chorea, unless one has an opportunity to observe the chronic progressive course later in the disease. The evidence of heredity is also diagnostic in the differentiation of this form of chorea, from senile chorea due to arteriosclerotic and degenerative processes of the cortex, although the latter begin rarely before the age of 55.

Oppenheim and Remak report a form of "familial chorea" in two boys, sons of a woman suffering from a chronic form of hemichorea. Both boys became affected at the age of 8 with a typical chronic, progressive chorea, which involved at first the lower extremities and gradually

spread to the entire body, with a predominance of symptoms in the legs.

Some authors regard Unverricht's myoclonia similar to hereditary chorea, but this, in the opinion of Oppenheim, is not justifiable.

The differentiation between Huntington's chorea and paresis, when the latter is associated with choreic movements, i.e., "choreatic paresis," may be very difficult. On the mere positive biological findings as to paresis, the diagnosis of Huntington's chorea cannot be excluded, because the two conditions may coexist (Lowrey and Smith³).

Association with Other Diseases.—Huntington's chorea may be associated with epilepsy, which may begin simultaneously with the chorea, or it may precede it by months or years. In one of Remak's cases epilepsy was present from the twenty-third to the thirty-first year and the chorea came at 40. Dieffendorf saw a case in which epilepsy developed at 17 and chorea at 53. In one of our cases at the Montefiore Home and Hospital the chorea began simultaneously with the epileptic convulsions.

Treatment.—**PROPHYLAXIS.**—It has been suggested by Muncie and others that, owing to the hereditary influences, the State concern itself with the investigation of the progeny of every case of Huntington's disease, and order sterilization of all those who already show symptoms, and to secure legislation that such of their offspring as show premonitory signs of the disease shall not be allowed to reproduce.

TREATMENT OF SYMPTOMS.—The treatment is purely symptomatic. Arsenic has no effect. The **bromids**, **hypodermatic injections of hyoscin**, **hydrobromate** and other sedatives in conjunction with **hydrotherapy** are employed for the relief of the twitchings, but the results do not seem to be very encouraging. In the treatment of these patients, it is important to take all precautions to prevent them from committing suicide.

Course, Duration and Prognosis.—The course of the disease is a very slow but progressive one; cases have been recorded whose duration was from one to two years (Gowers), but so short a duration is very rare. The disease is incurable and lasts several decades; it has very little tendency to shorten life. There are cases reported with recoveries, but it is doubtful whether they were cases of genuine Huntington's chorea. In so-called "choreatic paresis" if antiluetic treatment can be resorted to early and intensively, the prognosis should be comparatively favorable. Towards the end of the disease the patients become bed-ridden, and succumb to some intercurrent disease or cachexia from lack of nourishment, bed sores and coma. Very many patients become despondent and commit suicide.

Pathology and Pathogenesis.—The pathology of the condition is not at all clear. Charcot was unable to regard Huntington's chorea as a distinct clinical entity. Lannois and Paviot⁴ found proliferation of the neuroglia and infiltration of the cortex with glia cells as the chief changes, and they attributed to them a very important part in the origin of hereditary, familial nervous diseases. Kattwinkel found atrophy of the supratangential fibrous layer and the radiating fibers in the central convolutions with an accumulation of round cells around

the Betz* cells of the cortex. Rossi, Buck and Spiller found similar changes. Besta attaches great importance to the vascular changes.

Stier⁵ and Müller⁶ think the disease due to congenital malformations of the motor cortex, on the basis of which the subsequent changes develop. The former also found areas of diffuse degeneration of the spinal cord.

Oppenheim and Hoppe found disseminated miliary encephalitic foci in the motor cortex. Greppin, Kalischer, Kronthal, and Facklam found similar changes. Binswanger found changes which, in his opinion, did not essentially differ from those in paresis. Daniel J. McCarthy also found in three of his cases, which came to autopsy, changes not unlike those of paresis.

Raecke⁷ found the greatest evidence of disease in the cortex of the central convolutions, less marked in the occipital lobes, but there were no vascular changes in the sense of exudation.

J. Collins⁸ found in one of his cases the lesions to be primarily one of chronic parenchymatous degeneration of the motor cortex, and slight degeneration of the pyramidal tracts in the spinal cord.

Damaye⁹ found in two cases meningo-encephalitis with an intense neuronophagia in all stages. Kéraval-Raviot found in one case a meningo-encephalitis, and the smallest vessels completely enveloped by round cells just as in paresis. This, in their opinion, also accounts for the dementia.

Alzheimer¹⁰ found in a histopathological study of 3 cases, in addition to cortical changes, advanced degeneration in the cells of both the caudate and the lenticular nuclei; a marked diminution in the number of the ganglion cells, with a corresponding increase of the glia nuclei, without the formation of glia fibers. He also noted the presence of lipoid material in the ganglion cells and in the vessel walls. Evidences of cell degeneration were seen in the subthalamie region and, to a lesser degree, in the nuclei of the optic thalamus, the pons and the medulla.

Ballentine¹¹ found a general shrinkage of the cellular elements, with some increase of the lymphoid cells of the pia with pigment. The nuclei in the first layer of the cortex were moderately increased, as were the satellite cells, and cells of the same type were to be seen along the cortical blood-vessels. In another case, the same author found a large amount of cerebral fluid, 250 c.c. The weight of the brain was 950 grams. There were also thickening of the membranes and a marked shrinking of the frontal and parietal convolutions.

J. A. Pfeiffer¹² found in two cases slight atrophy with an increase of the spinal fluid. The meninges were somewhat thickened, otherwise normal. There was no dilatation nor ependymitis of the ventricles. The smallness of the brain was conspicuous in both cases. The degeneration of the nerve elements was most severe in the corpus striatum, optic thalamus, and frontal, precentral and postcentral regions. In the medullated fibers of the cortex, there was a deficiency of the tangential fibers, and the oblique fibers forming the interradiary and supraradiary plexus were similarly affected. The radial fibers of Meynert's radiation

were well preserved. The ganglion cells showed different types of degeneration; many of them were acutely degenerated, many more were sclerotic; they all had a great increase of lipoid pigment within them; most of the Betz cells were markedly well preserved. There was an enormous increase of the glia cells, glia fibers, especially in the lower layer of the cortex, corpora striata and the thalami, the cells having small pyknotic, darkly staining nuclei being the predominating type. The most obvious changes in the vessels were in the lenticular nuclei and the optic thalami.

The walls of the capillaries were thickened and their lumen sometimes obliterated. An extraordinary number of amyloid bodies were present in the posterior columns of the spinal cord, thalami and the lenticular nuclei.

E. V. Niessl-Mayendorf¹³ studied in great detail the brain of a woman 32 years of age who died from chronic chorea. There were found, in addition to the usual cortical changes, distinct alterations in the important tracts connecting the cortex with the cerebellum and in the associated nuclei of the cerebellum.

Margulis¹⁴ found proliferation of the glia in the cerebrum, bulb and cerebellum. There was atrophy of the parenchymatous elements in the cortex, especially of the nerve cells. First, the cortex was involved, then the subcortex, and finally the tangential and superradiating cortex fibers. He believes that the disease is a congenital degeneration—a chronic gliosis. The hyperkinesis he ascribes to the irritation of the parenchymatous elements. The process, in his opinion, is one of the whole central nervous system and cannot be localized.

P. Marie and J. Lhermitte¹⁵ found atrophy and degeneration of the cerebral cortex and corpus striatum. They regard the affection as a chronic toxic encephalitis, and would refer the intelligence defect to the cortical changes, and the choreiform manifestations to the alterations in the corpus striatum.

G. Kiesselbach¹⁶ found gliosis with occasional small areas of softening in all parts of the brain. He believes the corpus striatum to be the seat of coördinate movements, the lack of which produces chorea; the special involvement of the putamen and caudate nucleus is, in his opinion, of great significance.

Dunlap reported at a meeting of the Section of Nervous and Mental Diseases of the New York Academy of Medicine, held March 14, 1916, that in a large series of cases in addition to other changes he found uniformly an atrophy of the caudate nucleus and putamen, with degeneration and marked reduction in the number of ganglion cells and an increase of the nuclei in the glia. J. R. Hunt, who studied the same sections from five of Dunlap's cases, found the large cells of the globus pallidus throughout the corpus striatum well preserved amid the wholesale loss and destruction of the smaller type of ganglion cells of the neostriatum.

Jelgersma found atrophic changes in the head of the caudate nucleus

which was reduced to one-third of its usual size. In his specimens there was also a marked proliferation of the glia cells.

J. R. Hunt says: "There can be no doubt as to the constancy of the pathological changes in the corpus striatum in Huntington's chorea." He believes that the corpus striatum is the great infracortical center for the control and regulation of automatic and associated movements. It is composed of two cellular systems, viz., the small ganglion cells of the caudate nucleus and putamen (neostriatum) and the large motor cells of the globus pallidus. The function of the neostriatal cells is inhibitory and coördinating; that of the pallidal cells is motor. Loss of the small cell inhibitory system is followed by chorea; loss of the large cell motor system is followed by the paralysis agitans syndrome, viz., rigidity, tremor and disturbance of automatic and associated movements.

From this array of findings and opinions, it becomes evident that there is no anatomical basis for the disease. The most common pathological changes found were those of a chronic diffuse cortical encephalitis, atrophies, meningeal thickenings, vascular changes, etc., etc. Unfortunately the changes found are not characteristic, but are common to a number of nervous and mental conditions of adult and advanced life. The explanation for the symptoms of Huntington's chorea must be sought at the *site* of the lesion. From all the evidence adduced up to the present time, it seems that lesions of the basal ganglia might be held responsible for the manifestations of the disease.

Historical Summary.—The disease was first described in 1872 by George Huntington of Pomeroy, Ohio, at the time a practitioner on Long Island. It seems that the disease, as described by Huntington, has long been familiar to physicians practicing in districts adjacent to Long Island. Dunglison¹⁷ quotes a letter written to him by Dr. Walters of Franklin, New York, giving a description of the malady almost identical with that of Huntington; it was then known to the laity as "magrums." Lyons, in 1863, published an article on chronic hereditary chorea in the *American Medical Times*, in which he described the disease as we see it to-day. Gorman, in his inaugural dissertation, delivered before the faculty of the Jefferson Medical College, stated that the disease as described by Lyons was prevalent in certain parts of Pennsylvania.

Speaking of his personal experiences with this form of chorea (Huntington's chorea), Huntington,¹⁸ in an address delivered before the New York Neurological Society in 1910, said: "Over 50 years ago, in riding with my father on his professional rounds, I saw my first case of 'that disorder,' which was the way in which the natives always referred to the dreaded disease. I recall it as vividly as though it had occurred but yesterday. It made a most enduring impression on my boyish mind, an impression every detail of which I recall to-day, an impression which was the very first impulse to my choosing chorea as my virgin contribution to medical lore. Driving with my father through a wooded road leading from East Hampton, L. I., to Amagansett, L. I., we sud-

denly came upon two women, mother and daughter, both tall, thin, almost cadaverous, both bowing, twisting and grimacing. I stared in wonderment, almost in fear. What could it mean? My father paused to speak to them and we passed on. Then my Gamaliel-like instruction began; my medical education had its inception. From this point on my interest in the disease has never wholly ceased."

Since Huntington's description of the malady, many instances have been recorded practically all over Europe and in various parts of this country. No cases seem to have been reported from Turkey, South America and the West Indies. While negroes seem to be comparatively free from Sydenham's chorea, Huntington's chorea has been met with in this race, associated with other nervous diseases, especially with epilepsy.

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OTHER FORMS OF CHOREA

Chorea of pregnancy, p. 518—Chronic intermittent chorea, p. 518—Chronic perennial chorea, p. 519—Senile chorea, p. 519—Post-hemiplegic chorea, p. 519—Hemiballismus, p. 519—Prehemiplegic chorea, p. 519—Choreic poliomyelitis, p. 519—Hysterical chorea (Chorea major, Chorea magna), p. 519—Chorea natatoria—Chorea malleatoria, p. 520—Localized, isolated or partial chorea, p. 520—Tantism, tigretier, p. 520—Chorea of the diaphragm, p. 520—Bulboparalytic chorea, p. 520—Dubini's chorea, p. 521—Electric chorea, p. 521—Bergeron's chorea, p. 521—References, p. 521.

Chorea of Pregnancy.—The reader is referred to the section on Complications, under Sydenham's chorea, for a description of this form.

Chronic Intermittent Chorea (Oppenheim).—In this variety relapses

follow so quickly and the intervals, as the disease progresses, become so short that the patients may be said to have chronic chorea.

Chronic Perennial Chorea (Oppenheim).—This form lasts for years or for a whole lifetime; it is very rarely seen in children. Oppenheim also recognizes a *chorea adultorum permanens*, which is distinct from the hereditary or Huntington's chorea.

Senile Chorea.—This is a form of chorea appearing in old age, and is a permanent condition, often, though not always, associated with mental disturbances. Riesman has observed a case of senile hemichorea, in which recovery took place. Bischoff has reviewed all the published cases of this variety of chorea and found that in about 20 per cent. of them the disease ran a typical course of chorea minor.

Posthemiplegic Chorea.—This is not a very common type. The disease is met with in incomplete hemiplegias, the movements being more marked in the paralyzed arm than in the leg; the muscles of the face, palate or tongue rarely are involved. The choreiform twitchings may be associated with a typical intention tremor, athetoid and associated movements; voluntary effort or movement increases the twitchings, and in the cases in which they occur, the muscles are usually relaxed and hypotonic. Posthemiplegic chorea is commonly a hemichorea, and is due to lesions in the posterior part of the optic thalamus, the red nucleus and the region of the superior cerebellar peduncle. When the lemniscus participates in the pathological process, the choreiform movements are accompanied by severe pain, the so-called "central pain," a characteristic symptom in the "thalamic syndrome." Mitchell called these cases "painful choreas." Touche, Henschen, Greiff, Lauenstein, Edinger and others report such cases.

Hemiballismus.—Kussmaul has described under the name of "hemiballismus" a form of hemichorea in which the involuntary movements are rhythmical and suggest "throwing movements."

Prehemiplegic Chorea.—This is another form of chorea, rarely seen, which is due to organic brain disease. When it does occur, it is found to develop in slow hemorrhages into the optic thalamus and is accompanied by sensory disturbances.

Choreic Poliomyelitis.—Epidemic poliomyelitis may occasion choreic movements, the jerking keeping up for days or weeks. The movements may precede the development of the poliomyelitic paralysis, they may last throughout the entire acute stage of the disease, or they may only be transient. This choreic type of poliomyelitis is more common in monkeys inoculated with the virus than in man. These twitchings in poliomyelitis are not to be confused with the tremors seen in the period of invasion of poliomyelitis. In establishing a diagnosis of paralytic chorea, it is important to bear in mind the possibility of the case being one of choreic poliomyelitis.

Hysterical Chorea (Chorea Major; Chorea Magna).—Hysterical individuals are frequently subject to general spasmodic movements, which at times are not unlike those of true chorea. These movements are sudden, shock-like, rhythmical contractions, mostly noticeable in the hands

and fingers, and not as severe as those in true chorea. Sometimes the involvement is more extensive and the movements are more severe. This type has been described by the French school as "hysterical chorea," and by the German school as "chorea major"; Oppenheim considers "chorea magna" identical with severe convulsive attacks of hysteria, and to have no connection with chorea.

Chorea Natatoria—Chorea Malleatoria.—Chorea natatoria, in which the patients make movements with their hands, like in swimming, and *chorea malleatoria*, in which the movements are similar to those of beating a hammer on an anvil, may also be considered as belonging to this clinical group. These forms of chorea have been known to disappear suddenly after mental or severe physical excitement, and upon the sudden reëstablishment of the menstrual flow, which had previously been suppressed.

Localized Chorea—Isolated or Partial Chorea.—There are rare cases of *localized, isolated or partial* chorea, in which the choreiform twitchings are limited to the lips, tongue, pharynx or larynx, and the movements may be very severe and persistent. These are probably neuroses, in the nature of tics or habit spasms, and are discussed in this section only because they are constantly being reported in literature under the name of "Chorea."

Tarantism—Tigretier.—Tarantism and tigretier are described as forms of chorea, both being manifestations of hysteria. Tarantism is not known to-day. It is supposed to be due to the bite of the tarantula, and when the "disease" develops, periods of depression and stupidity first occur. When the sound of a musical instrument is heard, the patients leap into the air and indulge in the wildest form of dancing and shouting, which is continued until they drop from exhaustion. During the attacks, sexual excesses of all kinds are indulged in.

Tigretier is a disease of modern times, very closely resembling tarantism; in fact, both of these are, in many respects, like those forms of religious excitement which are accompanied with suspension of inhibitory control and disordered muscular movement. The "convulsionnaires" in France and the "jumpers" in Maine are good examples of this type of disorder.

Chorea of the Diaphragm.—Recurring pleurisy of the diaphragm and injuries to the chest are occasionally followed by rapid, rhythmical contractions of the diaphragm, with attacks of dyspnea and dry cough. The older writers described these recurrent attacks as "chorea of the diaphragm." In recent literature Simonin and Chavigny¹ describe two cases of this affection in two soldiers, in both of whom the disease dated from long before the war.

Bulboparalytic Chorea.—Charcot has pointed out that there are some cases of chorea in which the choreic spasms may disappear in the course of the disease, and a form of paralysis appear instead. Bruns² emphasized the frequency of these true paræses; he believes that, because in some of these cases mutism and dysphagia may persist for months, these symptoms cannot be explained by any marked choreic unrest

in the muscles of deglutition and phonation, but that they must be due to a real motor weakness, and he calls this the bulboparalytic syndrome of chorea. Because salivation is not an uncommon symptom in severe forms of chorea, as has been observed by Price,³ there seems to be additional evidence that there are some cases in which involvement of the bulb in chorea may be a predominating feature.

Dubini's Chorea.—Dubini described a form of chorea, observed in northern Italy, which may appear at any age. It begins with pains in the head, neck or back; this is followed by electric-like, rapid, short muscle spasms, involving first one arm and one side of the face, later the homolateral leg, and lastly the other side. These shocks may be accompanied by unilateral epileptiform seizures. As the disease advances, paralysis comes on in the affected extremities, which soon becomes general. The muscles become atrophied and their electrical irritability is diminished. This form of chorea is very painful; there is a marked hyperesthesia of the skin, the slightest touch of it producing the most violent contractions. The mind remains clear. High temperatures are common. After days, weeks or months, death ensues in coma or from cardiac failure. Very few cases recover. The disease is probably infectious in origin. On autopsy, meningo-encephalitis has been found.

Electric Chorea.—Henoch describes a form of chorea in which the muscular contractions differ from those in infectious chorea, in that they follow one another with lightning-like rapidity. The muscles of the shoulder and neck are most commonly involved. There is no change in the nutrition of the muscles, and there are no sensory disturbances. The prognosis is good, except that the contractions are so frequent and intense that the patients are incapacitated from work. Recovery takes place in a few days or weeks after the use of arsenic and hydrotherapy.

• **Bergeron's Chorea.**—Bergeron described a similar disease except that it affected delicate and anemic children.

Summary.—There is very little known of the etiology or pathology of these curious forms of so-called chorea. Some believe them to be manifestations of auto-intoxication, others of epilepsy or hysteria, and still others as forms of myoclonia.

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PARALYSIS AGITANS

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Synonyms.—Paralysis agitans, Parkinson's disease, Shaking palsy.

Definition.—Paralysis agitans is an incurable disease appearing usually in the fifth decade of life, and characterized by the gradual onset of tremor, muscular rigidity and weakness, giving rise to a peculiar attitude, gait and facial expression.

Etiology.—**FREQUENCY.**—The disease is said to be more rare in Germany than in America. Berger¹ found 37 cases among 6,000 patients with nervous disease. Putzel² saw, during eleven years, 30 cases among 4,600 patients at the Clinic for Nervous Diseases in the outpatient Department of Bellevue Hospital. König³ found in Siemmerling's Clinic in Kiel, from 1901-1913, 23 cases among 5,000 patients with nervous disease. During the year 1914 there were in the wards of the Montefiore Home and Hospital, 30 cases of paralysis agitans among 161 patients with nervous disease. (Owing to the method of selecting patients for admission to this hospital, these figures are of no statistical value as to frequency.)

AGE.—The disease usually begins after forty years of age. In Gowers' 115 cases, two-fifths began between fifty and sixty, and about one-fifth in each of the two decades between forty and fifty and between sixty and seventy; it appears almost twice as frequently between sixty and seventy, on account of the lesser number of persons living during that decade. Occasionally it begins between thirty and forty, rarely under thirty or over sixty-five years of age. Gowers' series included 2 males beginning at seventy-three, 1 female at seventy-four, 1 case at twenty-nine (his youngest case). Hadden saw a case develop at twenty-five, Buzzard at twenty-one, Duchenne at nineteen, Berger at seventeen, Ballet-Rose at fifteen, Lannois at twelve, Weil and Rouvillois at ten. J. Ramsay Hunt⁴ reported 4 cases, 1 of which began at thirteen, 1 at fifteen, 1 at twenty-six and 1 at thirty. (Two of these were from the Neurological Wards of Montefiore Home and Hospital.) J. S. Bury⁵ reports 2 cases in a brother and sister at eighteen. Clerici-Medea reports 1 at twenty-eight and 1 at twelve.

SEX.—The disease is more frequent in males than in females. Gowers found it in 73 males and 43 females; König, 12 males and 11 females; Mendel found both sexes equally affected; Putzel, 19 males and 11 females; Montefiore Home and Hospital, 17 males and 13 females.

RACE.—Judging from the reports the disease seems to be more prevalent in the Anglo-Saxon than in the German race. It is of great rarity in the negro race. Up to 1913, Burr⁶ saw only one case in a negro at

the Philadelphia General Hospital, no other case having ever been seen in that institution, or at the Philadelphia Infirmary, or at the University of Pennsylvania Hospital. It is said to be comparatively rare in Italy. Among Caucasians it seems that it is the most highly strung who are prone to the disease.

HEREDITARY INFLUENCES.—As a general rule, the influence of heredity in the development of the disease is not marked, although it may be a predisposing cause in many cases. Both Berger and Gowers found in 15 per cent. of their cases a history of the disease in more than one member of the family. Of Wollenberg's 19 patients 5 had a family history of mental and nervous disease. Oppenheim knew of one family in which two sisters developed paralysis agitans at an early age, a third becoming affected late in life with senile dementia with peculiar choreic movements of the tongue. In another one of his cases of paralysis agitans, a brother had the disease and a sister had bulbar palsy. He also saw one family in which three sisters had the disease. From his cases he is led to believe that the disease has a special tendency to appear in long-lived families. Clerici-Medea⁷ saw in one family 2 sisters with the disease, and in both it developed at an early age (*see under Age*). Bury (*see under Age*) is also of the opinion that certain families are more prone to it than others. Bonnhoeffer and Siehr saw 2 cases in one family. Lundborg⁸ reports 7 cases of paralysis agitans in three generations of the same lineage in one Swedish peasant family; besides these, 2 more cases existed in that family which were never diagnosed by a physician.

SOCIAL INFLUENCES.—Occupation, exposure to lead, brass and mercury, and one's station in life seem to have little or no influence on the development of paralysis agitans. Living in damp rooms, or prolonged exposure to wet and cold has been, in Putzel's cases, a most efficient etiological factor. Walz thinks the disease occurs only in individuals who have a deteriorated nervous system. J. M. Clarke⁹ suggests that the condition is one of the premature senescence-abiotrophy of certain neuronie systems, which may occur at any age.

In our cases at the Montefiore Home and Hospital the patients affected with paralysis agitans were persons of good emotional equilibrium. Bad habits, dissipation, sexual excesses, overindulgence in food, alcohol or tobacco seem to have been no determining factor in our cases.

ANXIETY, WORRY, EMOTIONAL EXCITEMENT, FRIGHT, GRIEF, ANGER.—Prominent among the predisposing causes mentioned by most writers are anxiety, worry, emotional excitement, fright, grief and anger. Several cases due to fright developed among the inhabitants of Metz and Strasburg during the Franco-Prussian war. Lorain¹⁰ reports the case of a seventeen-year-old girl frightened by the bursting of a shell in the cellar in which she had taken refuge during the siege of Paris; she immediately developed a tremor of the right arm which soon extended over the entire body, and at the end of five years she was still suffering from paralysis agitans. In the light of our experience in the late war, it

would be interesting to know whether this was a case of genuine paralysis agitans, or of what, nowadays, we call shell-shock or "war neurosis."

Gowers could find in only one-third of his cases a direct cause.

TRAUMA.—Trauma with its accompanying psychic effects is well recognized as a predisposing cause. Walz's¹¹ analysis showed, in 26 cases: general concussion in 6 cases; stabs and cuts in 7; burning and freezing in 1; sprains, fractures and twists in 4; and contusions in 8.

It was Charcot who first pointed out that physical injuries in general were frequently a predisposing cause in the production of the disease, and furthermore that it often began first in the part which was injured. Krafft-Ebing,¹² however, found in his series of 110 cases, only 7 cases in which trauma could be attributed as a cause. He believes the cause to be fatigue due to overstrain in the muscles, because in 50 out of 88 cases he found the disease had developed first in the right upper extremity, and this was the extremity utilized by the patients in the course of their daily tasks. This is the reason he thinks that unless trauma to the lower limb is the predisposing cause, the disease begins most commonly in the upper limbs.

Trauma may also determine the spread of the disease in a case which is already showing signs of it.

INFECTIOUS DISEASES.—Cases have been reported after typhoid, malaria, syphilis and the other infectious diseases, but it is doubtful whether these can be considered determining factors.

Symptomatology.—**CLINICAL HISTORY.**—*Mode of Onset.*—The usual onset of the disease is gradual. Prodromal symptoms, such as burning or rheumatic pains, weakness, numbness, girdle pains and local non-inflammatory swellings, frequently precede the characteristic symptoms of tremor and rigidity. These are sometimes erroneously considered manifestations of rheumatism. They are vague and wandering, their distribution more or less general, or localized in the limb or limbs, which later become affected with the disease. They may make their appearance years or months before the development of the other symptoms.

PHYSICAL FINDINGS.—*Tremor.*—Tremor is the first symptom of the disease in two-thirds of the cases; there are rare cases in which it is absent throughout the entire course of the disease—the so-called *paralysis agitans sine agitatione vel tremore*.

The tremor consists of slow, rhythmical oscillations, 4-7 per second, noticed chiefly in the distal ends of the upper extremities, the hands and fingers. The movements take the form of flexion and extension, abduction and adduction of the fingers; flexion and extension, pronation and supination of the hands. They are usually within narrow limits, the thumb and index finger merely rubbing together as in "rolling pills" or "counting coins." The movements in the hand and forearm are less limited, and may sometimes amount to true shaking. The tremor is slight at the beginning and increases with the progress of the disease; it lessens in frequency as it increases in amplitude, the fine tremor of the early stage being quicker than the coarser tremor of the later period.

The most characteristic feature of the tremor as pointed out by Parkinson is that *it continues during rest*. It is present when the patient is lying down or standing, whether his hands are supported or hanging by his side. The character of the tremor is remarkably *stereotyped*; the same movements of flexion and extension occur in the same tempo and with the same amplitude of oscillation all the time. It may from time to time increase or diminish in its intensity, but only for a second or two, after which it resumes its original character.

Voluntary active movements stop the tremor for a time; it ceases momentarily when the patient extends his hand or alternately opens and closes it. Momentary cessation of the tremor may be observed when he grasps an object or changes the position of his limbs, or when he is made to fix an object with his eyes. This temporary cessation is due to the general inhibiting effect of active movement.

During forced active movements or during slow but continuous movement, such as writing, the tremor persists or is increased; active movement associated with excitement also increases it. A patient with shaking palsy may be more comfortable when walking than when sitting or lying down and may be able to walk long distances without being annoyed by the tremor. The tremor ceases in the limbs involved in an attack of hemiplegia, to return with the recovery from the hemiplegia. Variations occurring in the rhythm or rapidity of the tremor later in the disease are due to the effect of the accompanying muscular rigidity. The relation between tremor and rigidity is aptly expressed by Hughlings Jackson when he says, "Tremor is rigidity spread thin, and rigidity is tremor run together."

Passive movements may stop the tremor for a time or even altogether (Oppenheim), but it is then to some extent transferred to other muscles and is increased in the extremity which is not being manipulated. Concentrating the patient's attention, or a mere attempt to touch the shaking limb, may also temporarily inhibit the tremor. The tremor always ceases during sleep and under narcosis.

The shaking may be confined to one arm or hand or it may involve both arms, or an arm and leg on the same side, or all four extremities. In the hemiplegic form the facial muscles may also participate in the involvement. Pronounced tremor is generally less common in the lower than in the upper extremities. When the legs are involved, the tremor is most noticeable in the calf muscles, but it may be seen in the thighs and even in the toes. Occasionally the muscles of the trunk and back may participate in the tremor, but it is almost never seen in the abdominal muscles. The head is not always spared; tremor of the lower jaw, of the muscles of the chin, lips and tongue is not uncommon. Gowers saw the tremor once in the orbicularis palpebrarum and Westphal in the lower facial muscles. In one of Oppenheim's cases the tremor of the eyelids was so intense that he could not make an ophthalmoscopic examination of the patient's eyes.

Rarely do the muscles of respiration participate in the tremor. Miller, Graeffner, Cisler and others report tremor in the vocal cords,

and Rosenberg saw it in the vocal cords and velum palati. (*Cf.* Speech, p. 527.)

Muscular Rigidity.—Another characteristic symptom is muscular rigidity; its intensity bears no relation to the intensity of the tremor. As a rule, it sets in later than the tremor, and the patients may be conscious of it long before it can be elicited by the examiner. The facial expression, attitude, gait, and the slowness in the execution of voluntary movements are all due to this rigidity.

The rigidity is "cadaveric" and differs from that of hemiplegia or paraplegia in that it is permanent and is not increased by the movement which elicits it. It produces a resistance in the limbs which does not vary, whether the limbs are moved slowly or rapidly. Passive movement elicits a slightly "interrupted" rigidity of the muscles—the so-called "cog-wheel" phenomenon (Moyer)—not unlike the sensation experienced by one on "pulling a ratchet." Another peculiarity is that, in spite of its severity, the rigidity may be overcome on repeated passive movement, except in the older cases in which marked contractures have already resulted.

Attitude and Facial Expression.—As the disease advances it produces almost absolute fixation of the entire body, causing the patient's head to be inclined forward, as if fixed to the trunk. The facial muscles, having lost their emotional play almost entirely, the face looks as though it were covered with a mask. The back is curved in kyphosis—rarely in lordosis or scoliosis. The patient holds himself "rod-like," with arms abducted, the elbows flexed and the wrists extended, with the fingers either flexed at all joints or at one joint; at times—especially when the interossei are involved—the fingers may be extended, or the hand may assume a position similar to that in which a pen is held (Charcot's writing hand), or the fingers are dug into the palm of the hand (Déjerine's fakir hand). The thighs are flexed and adducted, the knees bent and the feet held in a position of talipes equinovarus with a claw-like deformity of the toes.

Muscular Weakness.—Muscular weakness and rigidity usually come on together, and are as characteristic of the disease as the tremor. The loss of muscle power varies much in degree: at first it is slight; it gradually increases, being usually greatest in the part in which the tremor developed first and most. This weakness must not be confused with the general weakness seen occasionally as a prodromal symptom. It never amounts to a total paralysis; it may precede the tremor or may begin simultaneously with it. Impairment and retardation of active movements in the disease are due to the muscular weakness and the rigidity, although it may be noticed even before the rigidity is very marked. Active movements are slow; the muscles do not seem to respond immediately to the will. The simplest movements are carried out sluggishly, as if with great deliberation; the more complicated the movement, the greater the sluggishness.

Gait.—In a typical case of paralysis agitans, when the patient is asked to walk across the room, he begins hesitatingly, with short, shut-

fling steps, and hurries more and more until he reaches his destination—"festinating gait"; if at the point of destination he has no support or no object to grasp, he does not stop but falls to the ground. This is called "*propulsion*." The patient "runs after his center of gravity." There is sometimes a tendency to run backward—"retropulsion." Retropulsion occurs when the patient bends too far back, as in trying to take an object from a shelf. A similar phenomenon may be elicited when the patient is attempting to walk in a lateral direction—"lateropulsion." These peculiarities in gait can best be brought out by giving the patient a push in the desired direction or pulling his coat backward. One of our patients at the Montefiore Home and Hospital could walk much better backward than forward. Propulsion and retropulsion are much more common than lateropulsion. Oppenheim explains these disturbances in gait as due to the fact that the patient has special difficulty in bringing the groups of muscles which have been in a condition of rest or tonic spasm rapidly into one of contraction or relaxation. He cannot arrest the motion and is compelled to continue the movement in the direction once taken.

Loss of Associated Movements.—J. Ramsay Hunt lays great stress on the early loss of associated movements of the arms in walking. When a patient with paralysis agitans is asked to swing his arms voluntarily, he has no difficulty in doing so, no matter how much rigidity there may be in his muscles, but on walking, he holds his arms stiffly side by side, and the natural swing of the arms as seen in a normal individual is absent. An attempt at this "natural swing" of the arm can even be noticed in a patient with hemiplegia due to a lesion in the course of the pyramidal tract; careful examination of a patient of this kind will show that he cannot swing his arm voluntarily when asked to do so, but when walking, no matter how much spasticity or paralysis there may be in the arm, the tendency to swing it will be at once noticeable. The absence of associated movements in paralysis agitans can also be readily demonstrated when the patient is asked to make a "fist"; the normal extensor "kick" of the wrist seen in a normal or hemiplegic individual is entirely absent. The marked abduction of the thumb and the spreading of the fingers on opening the hand quickly, seen in a normal individual, are also absent in a patient with paralysis agitans. This loss of rhythmical associated movements of the arms in walking, on making a fist and opening the hand, constitutes, in Hunt's opinion, an important sign of the clinical types of "pallidal palsy." (Cf. Pathology and Pathogenesis, p. 537.) The author has found the loss of these associated movements a helpful aid in the differentiation of paralysis agitans from the functional as well as from the hemiplegic type of paralysis due to pyramidal tract involvement.

Speech.—The voice, in paralysis agitans, is weak and whining; speech does not begin until some time has elapsed after the thought has been created. There is no modulation in the voice; after the patient has once begun to speak, the words roll over each other. There is no scanning, and rarely is it explosive in nature. These changes in speech

depend upon the amount of tremor and rigidity in the muscles of the larynx and articulation.

The study of the larynx in paralysis agitans is difficult, on account of the presence of false tremors, which may be very misleading. As has been pointed out, true tremor may be present in the vocal cords, as in the other muscles of the body. Graeffner¹³ has shown that at times the tremor may be ipsilateral (on the same side) with the tremor of the upper extremity, or it may be contralateral (on the opposite side). In 80 cases he found 21 in which there was a tremor of the entire larynx of the same tempo as the tremor of the body; in 27 the tempo was different; in 32 there was no tremor of the true vocal cords. In 8 cases without tremor of the body, tremor of the larynx was observed in 5 cases. The tremor of the larynx, in all the cases in which it was present was most evident in the open position of the vocal cords. In 12 of the cases the adductors were involved. Cisler¹⁴ found in 75 per cent. of his cases a "cadaveric rigidity" of the cords. According to Glogau,¹⁵ a mouth record taken in cases of paralysis agitans will show from the impulse to speak to the first explosive word a straight line corresponding to the reaction time between the two.

Other speech disturbances, such as stammering, stuttering or aphasia have never been observed in this disease.

Eyes.—There is never a true paralysis of the ocular muscles, and there is no nystagmus. The ophthalmoscopic examination shows no changes in the fundi which can be considered characteristic of the disease. Oppenheim noted a paralysis of convergence in one case, which he thinks was due to a tonic contraction of both abducens muscles. Slowing of the eye movements and oculomotor paralysis have been described (Saint-Leger, Debove and Neumann). In two of Oppenheim's cases of unilateral paralysis agitans there were oculopupillary symptoms on the affected side; in another unilateral case he also noticed the presence of a von Graefe. Moczutowsky (cited by Oppenheim) lays stress on the fact that when the eyes are opened, the frontal muscles may sometimes be seen to be in a condition of tonic contraction, so that the folds on the forehead disappear gradually instead of suddenly. Oppenheim never observed this phenomenon.

Bulbar Symptoms.—Dysarthria, dysphagia, salivation and drooling have been noted. Bernhardt noted, in one case, forced laughter similar to that seen in pseudobulbar paralysis. Oppenheim once saw excessive salivation associated with an uncontrollable discharge of mucus from the nose.

Reflexes.—The superficial and deep reflexes are always present, rarely increased; later in the disease when the rigidity is marked they may be difficult to elicit. There is no Babinski or any of its modifications present. False clonus due to tremor may be observed. Oppenheim has been able to elicit true clonus in a few exceptional cases. Tileston¹⁶ also observed true clonus without other signs of pyramidal tract involvement. Graeffner has found the pharyngeal reflex absent in 20 out of 34 cases and markedly diminished in 9 cases. Mendel's observations

in reference to the loss or diminution of the Achilles reflex were not borne out in 28 cases examined by Graeffner, although this reflex was found to be modified in a number of the cases; these modifications, he believes, were more in conformity with the general findings in the senile and arteriosclerotic.

Huet-Alquier and others regard exaggeration of the deep reflexes as the usual condition. Our cases, in the absence of complications, never showed any abnormality in the reflexes.

Westphal's paradoxical reflex may often be seen, not only in the extensors of the foot, but also in the other muscles. This reflex is a contraction produced by the passive approximation of the origin and insertion of any muscle. If, for example, the patient's foot is pressed upward, a tonic contraction appears in the extensors of the foot, most noticeable in the tibialis anticus, which persists for a considerable time, and maintains the foot in a position of dorsiflexion. This symptom is of no value when the patient, in the belief that he should keep the foot in this position, *actively* contracts the extensor muscles.

Abnormal associated movements in the toes of the healthy foot, when the patient attempted to move the affected foot, have been seen by Oppenheim and Frank.

Drop-wrist or the "grasping reflex," which is a normal phenomenon in infants, but is inhibited in adults, appearing only when the higher cortical centers of inhibition are destroyed, has been noted by Bechterew in focal lesions of the cerebral hemispheres in the late stages of general paresis and diffuse arteriosclerosis accompanying hemiparesis. It has also been noticed by Janischewsky in one case of paralysis agitans. Bechterew thinks the center of this reflex is in the optic thalamus.

Sensation.—There are no objective sensory disturbances in this disease. Hyperesthesiæ, paresthesiæ and hypesthesiæ are common both as premonitory symptoms and throughout the course of the malady (*Cf.* Karplus¹⁷). Some patients complain of a feeling of excessive heat or cold or vertigo. The sensation of heat does not depend upon an elevation of the body temperature. Grasset and Apollinario have found a rise of temperature on the *external* surface of the body and Fuchs has observed an *actual* rise of temperature. A rise in the body temperature, in the majority of cases, is, however, an indication of some acute complication.

Acute pain, except as a premonitory symptom, is, as a rule, not found in paralysis agitans, although the French school has described a painful form of the disease which they call "*forme douloureuse*" (L'Hirondel). The patients seem to experience great trouble at night because, on account of their rigidity, they cannot turn around in bed to change their position, and in some cases the paresthesiæ may be so distressing that they cannot tolerate even the pressure of the bed clothes on their bodies.

Muscles.—The muscles show no change in size; late in the disease, when the patients have reached the stage of general helplessness, atrophy, due to disuse, may be seen.

As a general rule the electrical irritability of the muscles to both currents remains normal. Borgherini¹⁸ found a delay in the latent period of muscular contraction and a lessened irritability of the muscles and nerves to the electric current. The weight of authority is against these changes.

Skin.—Hyperidrosis is common. The skin of the hands may be smooth, erythematous and shining; occasionally it is edematous, known in French as "main succulente."

Joints.—The joints are usually not affected, although French writers have suggested that there might be true parkinsonian arthropathies. Oppenheim thinks that these joint affections are probably combinations of paralysis agitans with arthritis deformans.

Sphincters.—In typical cases there is no involvement of the sphincters. If the subject of the disease is well advanced in years there may be complications, such as a hypertrophied prostate in the male, or a prolapsus uteri in the female, which for mechanical reasons may produce rectal or bladder disturbances.

Gastro-intestinal Tract.—Owing to the sedentary habits and lack of exercise, most of these patients have poor appetites and suffer from constipation. Oppenheim observed, in one case, severe gastric disturbances with precordial pains and obstinate constipation one year before the development of the disease. Raymond records a case in which jaundice was a prominent symptom at the onset of the disease.

LABORATORY FINDINGS.—*Blood and Blood-Pressure.*—In uncomplicated cases there are no changes in the blood-picture or in the blood-pressure.

Urine.—Phosphaturia is common; the sulphates have been found decreased in amount. The urea nitrogen is normal.

Cerebrospinal Fluid.—The cerebrospinal fluid showed nothing pathological in any of the cases at the Montefiore Home and Hospital. Camp, of Ann Arbor, found in 10 of his cases that the globulin and albumin were not increased; the reducing substance was present, and the count did not average above four cells in any case.

SPECIAL FINDINGS.—*Psychic Symptoms.*—Parkinson, in his original essay, mentions no mental symptoms as constituting a part of the clinical picture. According to Ball (cited by Putzel¹⁹), paralysis agitans and insanity are associated more frequently than is commonly believed. König found many cases of hypochondriasis, melancholia and paranoid conditions among his patients; rarely, if ever, was euphoria present; in certain cases a true psychosis was developed; senile dementia and terminal deliria were frequent combinations.

In general, it may be said that the intelligence in most of the cases is unimpaired. The distressing symptoms toward the end, with their general helplessness, make the patients peevish, irritable and depressed. In our experience, no matter how severe the disease was before they had become bedridden, they all seemed to be good-natured and satisfied with their lot. By the weight of authority, mental symptoms early in the

disease are not a characteristic part of the clinical picture, and are to be considered complications rather than symptoms of it.

Diagnosis.—The slow, progressive development in the fifth decade of life of a chronic painless disease, characterized by tremor, muscular rigidity, a mask-like face, statuesque and rod-like station, with a festinating gait, is diagnostic of a typical case of paralysis agitans. There are, however, many atypical cases which may need prolonged observation before their true nature can be determined.

DIFFERENTIAL DIAGNOSIS.—*Multiple Sclerosis.*—Paralysis agitans is distinguished from multiple sclerosis by the fact that in the latter the tremor is coarser, not as rhythmical, and occurs only during the performance of a voluntary movement, i.e., an intention or "action" tremor, which is associated with nystagmus, optic nerve changes, a scanning speech, exaggerated reflexes, and bladder disturbances, and affects younger individuals. If the paralysis agitans begins with weakness and rigidity, without tremor, the typical attitude of the limbs, head and trunk will be diagnostic, but when the disease is unilateral, and without tremor, a correct diagnosis may be impossible.

Paresis.—In paresis the tremor is not rhythmical, and does not persist during rest. The absence of characteristic attitude and gait, with the mental changes and the positive biological findings in the blood and cerebrospinal fluid will make the diagnosis clear.

Senile Tremor.—In senile tremor the head is chiefly affected, and the tremor is brought on or intensified by active movements; there is no rigidity or peculiar attitude and gait. Senile tremor occurs much later in life than the usual cases of paralysis agitans.

Senile Arteriosclerosis of the Brain and Cord.—This condition may produce a clinical picture resembling paralysis agitans. The presence of true paralyzes, with bladder involvement, dysphagia or dysarthria, resembling pseudobulbar palsy in the cases of cerebral arteriosclerosis and exaggerated deep reflexes with a spinal type of sensory disturbances, when the cord is involved, will be diagnostic criteria.

Unilateral Spastic Hemiplegia.—Unilateral spastic hemiplegia of a slowly developing type may simulate unilateral paralysis agitans, but it will be differentiated from it by the presence of symptoms of pyramidal tract involvement.

Hysteria.—In hysteria the presence of hysterical sensory manifestations, and the mental condition of the patient, in addition to the character of the tremor which is irregular, of greater amplitude, with a tendency to appear in attacks influenced by suggestion and hypnotism, will aid in differentiating the two conditions.

Traumatic Neuroses.—Traumatic neuroses are sometimes followed by a tremor, attitude and gait similar to paralysis agitans, and inasmuch as genuine paralysis agitans frequently follows trauma, a perplexing diagnostic problem may arise. Restricted visual fields, disturbances of sensation, exaggerated reflexes, and lack of progression, with possibly a history of litigation for personal injuries, will be in favor of the diagnosis of a traumatic neurosis.

Chorea.—The jerky, coarse, irregular character of the movements of chorea can hardly be mistaken for shaking palsy.

Arthritis Deformans.—Spiller reports a case of paralysis agitans diagnosed as arthritis deformans, until a tremor appeared in one of the limbs.

Cerebellar Disease.—Cerebellar disease may give rise to lateropulsion or propulsion with tremor, but the coarseness of the tremor combined with ataxia, hypotonia, dysidiadokocinesia, dysmetria and asynergia, in cerebellar disease, will serve to distinguish between the two conditions.

Wilson's Disease.—The juvenile form of paralysis agitans may sometimes be confused with Wilson's disease (progressive bilateral lenticular degeneration) but the coarseness of the tremor, the early onset of dysarthria and dysphagia, with the characteristic mental symptoms, and the very rapid course in the acute cases with fever and emaciation, in the latter, will be of diagnostic significance.

Tremors of Metallic Poisoning.—The tremors seen in chronic metallic poisoning will be distinguished from the tremor of paralysis agitans, by the history of occupation or exposure, the mode of onset and the course of the disease. (For further differential diagnosis, see section on Tremors, p. 463.)

Complications.—Symptoms of focal disease of the brain and spinal cord due to arteriosclerosis may frequently be associated with paralysis agitans, but inasmuch as such focal lesions may in themselves produce a parkinsonian tremor with rigidity, it is at times impossible to ascertain whether the condition is merely symptomatic of paralysis agitans, or whether it is a complication.

Berger has seen sudden attacks of *transient hemiplegic* weakness, and Gowers has known of occasional sudden attacks of *transient general powerlessness* occurring in paralysis agitans without any lesions being found in the brain to explain them. These are probably due to temporary spasm of the cerebral arteries—"intermittent cerebral claudication."

Early mental failure with loss of memory may be a complication of the disease. Gowers has once met with convulsions resembling those of *epilepsy* as a complication in a woman 59 years old in whom the convulsions and the paralysis agitans began at the same time. Buzzard has reported a case in which there was a *semi-cataleptoid* condition of the limbs. The combination with *tabes dorsalis* is very rare, as is that with *Basedow's disease*. Luzzato and Lundborg have seen paralysis agitans associated with symptoms of *myxedema*. Osnato²⁰ reports a case of paralysis agitans and *myopathy* occurring in an uncle and nephew, with evidences of internal glandular disturbances in the latter. J. Roux²¹ saw a man of 71 years of age with paralysis agitans complicated with an *acquired myotonia*. *Diabetes mellitus* was a serious complication in one of our cases of the disease.

Clinical Forms.—There is a tendency in modern medicine to consider paralysis agitans as being not a distinct clinical entity, but a syndrome with a definite characteristic complex of symptoms which may be due

to various pathological conditions. With this view in mind, J. Ramsay Hunt,²² of New York, recognizes three types of the disease: (1) the presenile and senile; (2) the symptomatic type; and (3) the rare juvenile form. Paralysis agitans without tremor or without rigidity and all the other various irregular and incomplete manifestations of the disease, the so-called "*formes frustes*," he considers subdivisions corresponding to variations in the symptomatology. He believes that all these are forms belonging to the paralysis agitans group, and that they are related to each other clinically because they all present the chief symptoms of the syndrome (rigidity and tremor) but that they present differences which are due to different localization and different kinds of lesions.

The juvenile type of paralysis agitans he considers a pure system disease, due to a slowly progressive atrophy of the motor neurons of the globus pallidus mechanism, while the presenile, senile and symptomatic forms are dependent upon senile and vascular changes in the course of the same mechanism.

JUVENILE TYPE.—The juvenile type is characterized by the unusual frequency of familial incidence, the early age of onset, the more rapid progression of the symptoms, the comparatively early involvement of the bulbar muscles, the rarity of the form without tremor and the infrequency of subjective sensory symptoms.

HEMIPLEGIC TYPE.—The hemiplegic type is not very common. In many of the cases reported as belonging to this type there was merely a predominance of symptoms on one side, but the facial expression and attitude were characteristic of typical cases of the disease. Juarros²³ reports a case in which the tremor and rigidity were exactly confined to one side of the body and there was no sensation of heat or perspiration on that side; the reflexes were normal. When the rigidity precedes the tremor in these unilateral cases, they may simulate a case of slowly developing hemiplegia; the same is true when the involvement in one limb alone persists for any length of time. Marshall Hall distinguished a *hemiplegic* and a *paraplegic* type and Berger added a *monoplegic* form. Gowers thinks that these should not be considered types, that they are merely prolonged stages of the disease, which later have a tendency to become generalized.

PARALYSIS AGITANS SINE TREMORE.—This is not a very uncommon variety. The tremor may be very slight or entirely absent; in these cases the diagnosis depends upon the rigidity, facies and attitude.

TYPES WITH VARIATIONS IN THE TREMOR—FORMES FRUSTES.—In some cases the tremor may be marked and constant in one part of the body, and be brought out in another part only on movement. In the latter cases the tremor may simulate that of multiple sclerosis (*see* Diagnosis, p. 531).

There are also rare cases in which tremor is present and rigidity slight or absent; in these, unless the tremor is very marked and typical, the condition may be impossible to distinguish from essential tremor, hysteria, or any of the other neuroses.

PARALYSIS AGITANS WITH HYPOTONIA.—Förster²⁴ described a form of paralysis agitans with hypotonia, i.e., tremor without rigidity, and Krämer²⁵ saw a rare "paralytic" type of paralysis agitans—typical paralytic disturbances without tremor and without rigidity. Hunt thinks that Krämer's and Förster's cases are vascular in type and not a system disease, not unlike the flaccid hemiplegias.

TYPES WITH PECULIAR ATTITUDES.—This type is not very common; in some the head may be inclined backward, or to one side, as in torticollis, or rarely the entire body may be in extension instead of flexion (Charcot).

FORME DOULOUREUSE, FORME RHEUMATISMALE.—In these cases pain of a dull character over the joints or limbs may be so marked as to be the predominating symptom in the clinical picture.

SYMPTOMATIC FORMS.—Paralysis agitans occasionally follows true hemiplegia, being limited to the limbs first paralyzed. It is questionable whether these cases can be considered true paralysis agitans, or complications of hemiplegia in which the tremor resembles paralysis agitans. This form has been noticed in lesions of the cerebral peduncles and of the midbrain, due to neoplasms, inflammation and arteriosclerosis.

The author recently had an opportunity of observing a case belonging to this group in the service of Doctor Strauss at the Montefiore Home and Hospital. A man of 43 years of age was admitted, complaining of fainting spells, weakness, and poor memory, all of which set in three years ago, following the death of one of his daughters. His previous history was absolutely negative. The only positive findings on examination were slight parkinsonian tremor with rigidity of the left arm and leg, with general features of hyperthyroidism which led us to diagnose paralysis agitans. Cranial nerves, cerebrospinal fluid, blood, etc., were negative. He had been going around the various hospitals and clinics in which his condition was diagnosed as hysteria. After three months' stay at the Montefiore Home and Hospital, without any previous acute disease or complaint, he became comatose and died a few hours later. Necropsy showed a large glioma involving the entire right temporal lobe, which had flattened out the lenticular nucleus, caudate nucleus and optic thalamus on that side without invading these, as far as could macroscopically be determined. The right ventricle was also partially collapsed and the septum lucidum of the third ventricle was pushed to the left.

ATYPICAL FORMS.—Many atypical and unusual cases of paralysis agitans, which it is impossible to classify, have recently been reported in medical literature.

The following cases are cited as illustrations:

Case 1.—Gilpin,²⁶ of Philadelphia, showed before the Philadelphia Neurological Society (see transactions of meeting held on March 24, 1916) a man 50 years of age who had a synchronous tremor of the right hand and head, with a fixed countenance and awkward speech; his pupils were dilated and immobile; he also had a paroxysmal cough of a suffocative character, without laryngeal abnormality, except for a

slight displacement of the epiglottis. The tongue when protruded was slightly deviated to the left, and the left side of the face appeared more active than the right.

Case 2.—Janischewsky²⁷ reports a case of paralysis agitans in a man who had difficulty in voluntary movements of the face and tongue, but who could fix his eyes on an object and keep them there in spite of the movements of the head; he could also follow the movement of an object with his eyes, no matter how rapidly it was moved. The author explains these phenomena as due to a lesion somewhere between the primary or lower centers for ocular movements and the cerebral cortex, the difficulty in movement and the rigidity in paralysis agitans being due, not to a paralysis in the sense of difficulty of voluntary contraction, but rather to a difficulty in relaxing the antagonist.

Case 3.—Vasiliu and Parhon²⁸ observed in a soldier, after an injury to the left parietal region, a parkinsonian tremor confined to the right middle, ring and little finger, with sensory changes. Operation showed only a fracture of the inner table, without any lesion in the meninges. In a few days after the operation the tremor disappeared and paralysis set in. These authors raise the question whether the tremor of paralysis agitans could not be cortical in origin.

Treatment.—GENERAL.—The disease being incurable, the treatment is necessarily **symptomatic**. The patient is to be maintained in good general condition; mental and physical excitement are to be avoided; a carefully selected diet, very light exercise and mild hydrotherapeutic measures are recommended. In the later stages of the disease the patient should be kept in as comfortable a position as possible, and all measures should be employed to prevent contractures. Charcot pointed out that these patients usually feel better driving in a carriage or in a railway train; this has led to the construction of "fateuils trepidants," chairs which, by their continuous oscillations, give ease and comfort.

ELECTROTHERAPY.—Electrotherapy is of little service. Some claim that a weak faradic current, frequently interrupted, diminishes the tremor. Oppenheim recommends the use of electric baths and gentle passive movements.

GYMNASTIC EXERCISES.—Gymnastic exercises to relax the rigid muscles allowing the various members of the body to fall against gravity are recommended by Friedländer.²⁹ During these exercises overstrain must be avoided. W. B. Swift,³⁰ of Boston, reports improvement after graduated slow exercise given to the arms, hands, legs and toes for fifteen minutes, three times a day.

HYDROTHERAPY.—**Warm baths** are useful to control the rigidity. Several patients at the Montefiore Home and Hospital, in the later stages of the disease, who had been rendered entirely helpless by the rigidity, were kept for two to three hours in a continuous bath of body temperature, and their pulse and blood-pressure taken every hour while in the bath. It was remarkable to see what beneficial effect this treatment had on the rigidity and tremor; some of the patients who

had to be fed for months before this treatment was instituted could feed themselves after staying in these baths for several hours. The improvement, however, was only temporary.

MEDICINAL TREATMENT.—**Bromids** alleviate the restlessness and the feeling of anxiety; **veratrum viride** and **gelsemium** are recommended by some to lessen the tremor, as are **cannabis indica**, **codein**, **opium**, **atropin**, **duboisin**, **scopolamin**, and other sedatives. Erb recommends **arsenic**, but in the hands of others it has been found of no value. The drug most commonly used and with the best temporary results is **hyoscin hydrobromate**; this may be given in fresh preparations by mouth, or preferably subcutaneously; during prolonged administration its toxic effects must be borne in mind although it seems that patients with this disease can tolerate unusually large doses, and can take it for a considerable period, without any marked ill effects. J. S. Bury³¹ reports two cases of the disease treated with **hyoscin**, 1/150 grain (0.0004 gram) twice a day, increasing it gradually to 1/96 grain (0.00067 gram) twice a day; during the two periods that the drug was administered, the tremor, the restlessness and the hot flushes were arrested.

GLANDULAR EXTRACTS.—**Thyroid extract**, as suggested by Lundborg, seems to have no effect. **Parathyroid preparations** have been used by various clinicians with varying success. W. N. Berkeley³² uses an acetic acid extract of fresh bullock's parathyroid glands, made by treating the triturated glands with cold distilled water, filtering, and then precipitating it with a very minute amount of acetic acid. Prepared in this way, it may be used hypodermatically in 15 minim doses, without the slightest irritation to the skin. He reports that 60 per cent. to 70 per cent. of the cases treated with this remedy for a period of six months were greatly benefited, and in them the progress of the disease was materially retarded.

Dercum³³ treated a woman of 57 years, having paralysis agitans, with 1/20 grain (0.00324 gram) parathyroid extract three times a day. She began to improve in a few days and was completely cured in a month. In this case the calcium metabolism was carefully investigated, and it was found that the calcium output was not influenced in the slightest degree by the administration of the parathyroids.

Pituitary extract has given Parhon-Urechí³⁴ good results. Gordon, of Philadelphia, claims that his patients were relieved by the administration of **calcium lactate**.

In our experience, the administration of the glandular extracts was followed by no results which we could not obtain from the administration of **hyoscin** and the use of continuous warm baths.

Course and Prognosis.—The course of the disease is progressive, slow and chronic; it may take from fifteen to twenty years before the patient becomes so bent that he must be confined to bed. The tremor may for many years be limited to one extremity, usually the arm. It may take from one to three years before the leg is involved on the same side as the arm; or the leg and arm may become affected simultaneously. More rapid extension of the disease is not very common. There is usually

a gradation in the severity of the tremor and rigidity in the different parts of the body, proportionate to the duration of the disease; there are, however, so many variations in the order of extension that it is impossible to foretell the future of a given case.

The prognosis as to life is good. Remissions may occur in the early periods of the disease, and may occur without as well as with treatment. Sometimes a remission is followed by an aggravation of the disease.

Owing to the fact that patients subject to this disease are well advanced in years, attacks of cerebral hemorrhage followed by paralysis frequently occur in the course of it. Parkinson, Westphal and Bychowski have noticed that attacks of apoplexy followed by hemiplegia led to a cessation of the shaking in the paralyzed limb, but it generally reappears. Collet, cited by Oppenheim, noted a case in which the tremor ceased suddenly on one side of the body, although no paralysis had appeared. The tremor has also been observed to cease before death.

The disease is *incurable*. French clinicians claim that the prognosis in the "rheumatic" form is comparatively good.

The disease may last from 10 to 30 or more years. Death is usually due to some intercurrent complication, decubitus (bedsore) or pneumonia. If no intercurrent affection sets in, the disease progresses until complete physical disability occurs and the patients, becoming bedridden, succumb to infection from bedsores and general cachexia.

Pathology and Pathogenesis.—Parkinson found an induration of the pons, medulla and cord. Charcot, Westphal, Berger and others could find no lesion in the central nervous system.

Dubief believes the anatomical basis of the disease to be lesions of "cerebrospinal senility." Hughlings Jackson thought the cerebellum was the part affected. Osler believes it to be due to changes in the cerebral cortex; Souques is of the same opinion. Brissaud regards the locus niger as the seat of the lesion. G. Maillard attributes the condition to general arteriosclerotic changes in the red nucleus.

Alquier found small disintegrated areas in the brain, but the motor area was well preserved in most of his cases. Haskovec and Barta found, in the brain, changes in the neuroglia cells, nuclear defects, pyknomorphia, rarefaction of the cytoplasm and vacuolization—changes more advanced than the ages of their cases warrant. There were also slight sclerotic changes in the vessels, hyaline degeneration in the capillaries and a marked proliferation of neuroglia along the axis cylinders in the peripheral nerves.

Spielmeyer thinks that the neuroglia cells which are similar to ameboid cells, and which he found in 6 cases in the white substance of the central nervous system play an important rôle in the pathology of the disease. F. H. Levy, however, suggests that these ameboid neuroglia cells are simply postmortem changes.

Borgherini found a dilatation of the perivascular lymph spaces, and a thickening of the vessel walls in the cerebellum with the same vascular

changes, overgrowth of the glia and pigmentation of the glia cells in the medulla oblongata.

In 1908, on purely theoretical grounds, Kleist and Zingerle suggested the region of the lenticular nucleus as the possible site of the lesion. Winkler described a loss of fibers in the lateral nucleus of the thalamus, in the inner limb of the lenticular nucleus, in the subthalamic region, in the tegmentum and in the pons. Before Kleist and Zingerle's suggestion, Manschot (Amsterdam, 1903—F. van Rossen) found a loss of fibers and cells in the thalamus, most marked in the lateral nucleus and atrophy of the putamen and subthalamic region. At about the same time, Jelgersma³⁵ found a reduction both in the size and number of the radial fibers in the lenticular nucleus, most marked in the globus pallidus; the strio-luysian fibers were atrophied, and the ansa lenticularis, ansa peduncularis and the H₁ and H₂ bundles of Forel were scarcely recognizable. There were similar changes in the lateral nucleus of the thalamus with atrophy of the superior cerebellar peduncles and the region between them.

F. H. Levy³⁶ observed similar lesions, except that the changes in the lenticular nucleus were more marked, and the dorsal nucleus of the vagus was also involved. In many of his cases there were lymphocytic infiltrations in the inner half of the globus pallidus and the paraventricular zone of the thalamus. He also called attention to the clinical resemblance of paralysis agitans, with the onset at thirty or forty, to the progressive degeneration of the lenticular nuclei as described by Kinnier Wilson.

Cadwalader found scattered areas of softening in the lenticular nucleus on each side; these softenings varied in size and were more numerous in the putamen than in the globus pallidus; the large nerve cells of the putamen were less numerous than normally. He found no involvement of the internal capsule, optic thalamus and external capsule.

M. Löwy³⁷ found symmetrical areas of softening in the lenticular and caudate nuclei in cases of paralysis agitans without tremor.

Jelliffe finds in paralysis agitans a paucity of the fibers of the internal capsule, especially of the groups of fibers derived from the cerebellorubrospinal tract. He regards paralysis agitans as a syndrome and not as a clinical entity.

Mingazzini³⁸ cites the case of a man who for four years had a paralysis agitans-like tremor in the right arm, with dysarthria and paresis of the right lower facial, partial sensory aphasia and dementia. Autopsy showed a sharply outlined loss of substance in the right caudate nucleus, a partial loss of the anterior segment of the internal capsule and the lenticular nucleus, with changes in the crural region of the cortex, in various parts of the brain stem and cord. In this case there was an almost complete lack of crossing of the right pyramidal tract, accounting for the symptoms on the same side as the lesion. Mingazzini's general conclusion is that such disturbances may produce symptoms resembling those of paralysis agitans.

E. W. Auer and S. P. McCough³⁹ studied pathologically two cases of paralysis agitans. They found in both (1) the corpus striatum to contain numerous small areas of rarefaction, giving the tissue a moth-eaten appearance; (2) clean punched-out holes—possibly enlarged perivascular spaces, from which the vessels may have dropped out (like in Wilson's disease); (3) round and oval basic staining deposits in the perivascular spaces and adjacent tissues (similar to those described by F. H. Levy); (4) a diminution in the number of the external medullary lamina and of the radial fibers of the lenticular nucleus with some evidence of degeneration of the latter (similar to those described by Jelgersma, F. H. Levy and others); (5) failure of the cells of the corpus striatum to stain well (possibly due to the age of the material). In one case, advanced degeneration of the cells of the centrum medium on both sides of the thalamus and subthalamic body was found.

J. Ramsay Hunt,⁴⁰ from a clinical study of four cases of juvenile paralysis agitans—one of which came to autopsy—concludes that paralysis agitans is a syndrome having a more or less common symptomatology, but including a variety of distinct pathological conditions, e.g., system degeneration (abiotrophy), senile degeneration with atrophy, vascular and perivascular lesions, toxic degeneration, tumors, lues, inflammation, etc., and that juvenile paralysis agitans is a pure system disease, characterized by a progressive primary atrophy of the efferent motor neurons of the globus pallidus system. There is an atrophy and diminution in the number of motor cells of the globus pallidus proper, the basal ganglion of Meynert, and especially the cells scattered through the caudate nucleus and the putamen (neostriatum). The atrophy was found to be most marked in the *large* cells of the neostriatum. In addition to the atrophy and disappearance of the ganglion cells there is a corresponding increase in the glia nuclei and a thinning of the fibers of the striohypothalamic radiations. The *small* ganglion cells of the neostriatum remain intact; the pyramidal tracts show no signs of atrophy or degeneration. The cells of the nucleus ruber, corpus Luysii and substantia nigra are normal.

Numerous investigators have found in cases of paralysis agitans an excess of neuroglia in the spinal cord, but they do not seem to agree as to its distribution or significance. Those who believe the disease to be due to lesions of the brain insist that the changes in the cord are accidental, or due to senility and arteriosclerosis.

Redlich⁴¹ found most of the glial tissue around the vessels in the anterolateral columns. Sanders⁴² found neuroglial proliferation more marked in the gray matter than in the white; in addition to the great amount of neuroglial tissue in the anterior horn, there were also numerous spindle cells present.

Dana found a moderate increase of connective tissue within the cord, and changes in the dendrites of the anterior horn cells, with a diminution in their number. Camp made a careful examination of the nervous system in fourteen cases of paralysis agitans with the Marchi method, but could not corroborate any of these findings.

Changes similar to those found by Dana have been described in Clarke's column and in the Betz cells of the cortex, the cell changes consisting of atrophies, pigmentation, lipoid degeneration, chromatolysis, vacuolization and tumefaction.

Vascular changes, such as miliary aneurysms, varicosities and arteriosclerosis of the finer capillaries in the cord, have been found by some, while others have ascribed the disease as due to occlusion or narrowing of the central canal, to amyloid bodies within the cord and various tract changes.

Ratner⁴³ describes fibrosis of the spinal cord around the septum and in the various tracts. His cases also showed in the gray matter an increased number of glia cells and degenerated ganglion cells; the tangential fibers in the region of the central fissure of the cerebrum were diminished in number, and the *ulnar nerve* showed pallor of the medullary sheaths with loss of the axis cylinders.

As far as our knowledge of the pathological anatomy of the disease is concerned, the statement made by Oppenheim ten years ago still holds true. He says: "We know nothing definite of the pathological anatomy of the disease. In most cases no changes were found. Virchow, Leyden and others found localized lesions, such as tumors of the optic thalamus and in the adjacent regions of the brain; but in these cases the paralysis agitans was symptomatic only; it was a post-hemiplegic type of paralysis agitans which must be distinguished from true paralysis agitans."

Inasmuch as the predominating clinical signs of the disease are referable to the muscles, the pathological basis for the disease was thought to be due to anatomical changes within the muscles. In fact, as far back as 1862, Skoda described fatty degeneration of the muscle fibers in Parkinson's disease. Gautier, Blocq, Sass, Schwerin and others regarded the disease as of myopathic origin; their proof, however, was not convincing. Catola and Salaris also claim to have found muscle changes. Buck, Demoor and Kinichi-Naka could find no such changes. Schiefferdecker-Schultze described lacunar atrophy and an increased number of small nuclei in the muscle spindles which, in their opinion, is the cause of the disease. Idelsohn emphasized the unusual prominence of the longitudinal striations of the primary muscle fibers, but he himself raised the point that this may be the result of unusual muscular activity. Oppenheim, who saw Idelsohn's specimens, is of the same opinion.

Camp, in studying the muscles in nine cases of the disease, found many fibers on cross-section swollen and round, instead of polygonal in shape, with a marked increase in the number and a peculiar chain-like arrangement of the nuclei within the fibers; in other parts of the same muscle there was atrophy of the fibers, with an overgrowth of connective tissue. In some of the cases the fibers had a hyaloid appearance, and the longitudinal striation was well marked, with a tendency to longitudinal cleavage. In one case he found an interstitial myositis with a trichina spiralis; in none of the cases could muscle

spindles be easily demonstrated. The nerve fibers within the "spindles" were normal. In this study Camp pointed out the significant fact that the changes were not of the same intensity in all the muscles nor in different parts of the same muscle.

Other investigators are inclined to attribute the pathogenesis of the disease to dysfunction of the ductless glands. Inasmuch as disease of the thyroid gland gives rise to tremor, the attention of investigators was naturally focused on this gland. In most cases no pathological changes were found. When it became recognized that the thyroid bears some relation to the adrenals, these too were studied, but no relationship could be established between the thyroid, adrenals and paralysis agitans.

Normal parathyroids being apparently the regulators of neuromuscular activity, Lundborg⁴⁴ advanced the hypothesis that paralysis agitans might be due to disease of these glands. Berkeley⁴⁵ was of the same opinion. Gobilovici⁴⁶ is also in favor of the theory that some relation exists between paralysis agitans, the thyroid, and parathyroid glands. Lundborg's hypothesis was strengthened by the researches of Loeb, J. B. MacCallum, W. G. MacCallum and C. Voegtlin on the relationship between the various muscular twitchings, calcium metabolism and parathyroids, and the improvements in tetany which followed the administration of calcium salts.

Morel, as a result of his experiences with autoplasmic grafts with parathyroids in tetany, also believes that the disease might be attributed to dysfunction of these glands.

Camp examined the parathyroids in two cases of paralysis agitans and found fatty perivascular infiltrations within them. Roussy and Alquier found these glands affected in several cases of this disease. Greenwald⁴⁷ argues that if the symptoms of paralysis agitans are due to parathyroid insufficiency, one might expect to find that the amount of acid soluble phosphorus in the blood serum of such patients to be greater than that of other individuals. In his experiments on the material at the Montefiore Home and Hospital, he did not find this to be the case.

Harbitz,⁴⁸ on finding at necropsy, in a case of paralysis agitans, symmetrically situated adenomata of the parathyroids, which gave clinically no local symptoms, thought that the presence of these produced pressure on the glands, with a resulting perversion in their secretions, to which he is inclined to attribute the disease. Manthos⁴⁹ also found the parathyroids much enlarged in a typical case of the disease.

R. L. Thomson⁵⁰ compared the parathyroids of 9 cases of paralysis agitans with those of 39 individuals, who died from other causes, and he could find nothing which could distinguish the parathyroids of the paralysis agitans cases from the others.

In this connection it may also be of interest to note that Parhon⁵¹ and Goldstein found changes in the hypophysis of a woman who had died of paralysis agitans.

Conclusions.—From the evidence presented thus far, it seems that no characteristic lesions other than those of the intercurrent affections

from which the patients died have been found with any degree of consistency, so as to lead one to say definitely what the pathology of the condition may be. The similarity between certain features of paralysis agitans and certain features of senility suggest that the disease may depend upon a premature senescence of certain regions of the brain, and that in some instances, owing to a hereditary predisposition, such processes may be more rapid, and may come on earlier than in others.

In a number of cases no lesions could be found, and this led some to regard the disease as a neurosis. In the majority of cases, however, lesions have been found in the brain, cord, peripheral nerves, muscles and ductless glands. These were not constant, and in the interpretation of their significance the question arose whether these lesions were primary or secondary, and due to senile and vascular changes. In fact, as one reads the clinical histories of the cases with the anatomical findings, his doubts are in many instances aroused as to whether they were genuine cases of paralysis agitans or only symptoms of organic brain disease, and, therefore, of secondary significance as far as the pathogenesis of the disease is concerned.

Thus in spite of the great amount of work done in pathological research, the pathology and pathogenesis of this disease still remain one of the most obscure chapters in medicine.

Historical Summary.—The disease was first fully described by James Parkinson, when he wrote his celebrated essay, in 1817, on the "shaking palsy." (An Essay on the Shaking Palsy, London, 1817). He differentiated it from the choreas, and Ordenstein, under the direction of Charcot, distinguished it from multiple sclerosis. The name "paralysis agitans" has been objected to by Charcot, because in some cases the shaking or the weakness may be slight or may not occur until late in the disease; still the name is undoubtedly most appropriate, because in the majority of cases both the shaking and the palsy are the most conspicuous phenomena.

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WILSON'S DISEASE

History and definition, p. 543—Symptomatology, p. 544—Physical symptoms, p. 544—Mental symptoms, p. 544—Clinical types, p. 545—Treatment, p. 545—Pathology, p. 545—Bibliography, p. 546.

Synonyms.—Bilateral progressive lenticular degeneration.

History and Definition.—In 1912, S. A. K. Wilson¹ described a peculiar disease, familial but not hereditary in type, occurring usually in individuals toward the latter part of the second decade of life, pursuing an uninterrupted chronic progressive course, varying in duration from a few months to several years, and terminating fatally. The disease is characterized clinically by tremor, rigidity, dysarthria and progressive

mental deterioration, and pathologically, by bilateral progressive lenticular degeneration, and a peculiar cirrhosis of the liver.

Symptomatology.—The most prominent physical features of the disease are:

PHYSICAL SYMPTOMS.—1. *Tremor.*—This motor disturbance is present in every case, although it may vary in intensity in different cases. At the onset of the disease, the tremor is fine, limited in range, and affecting only the distal ends of the extremities. It is regular, rhythmical, from 4-8 oscillations per second, increased by physical or mental exertion, and can be voluntarily inhibited for a brief period. As the disease advances, the tremor becomes more extensive, and toward the end, involves the entire body. The tremor is of the paralysis agitans type, with adventitious movements during active, voluntary efforts, very similar to chorea. Gowers saw such cases, and he called them "tetanoid chorea." In most of the cases reported, it appeared first in the right hand and was most noticeable when the patients attempted to write, so that bad handwriting came to be considered one of the earliest symptoms of the disease. Nearly as often it appears in the arms and legs simultaneously, less frequently in both arms alone, and in a few cases in the tongue.

2. *Rigidity.*—Another constant feature of the disease is the rigidity or spasticity of the muscles. It is progressive in nature, and involves all the muscles of the body except those of the eyes. As the disease advances, the rigidity gives rise to contractures, which eventually lead to permanent deformities. These contractures and deformities are best seen in the fingers, hands, toes and feet. The face, like in paralysis agitans, assumes a fixed and immobile expression. At times the contractures of the facial muscles of expression leads to an abduction of the angles of the mouth, and a separation of the lips, giving the patient a silly, almost idiotic appearance. There is nothing in the attitude of these patients which is more characteristic of this than of a number of other similar conditions.

3. *Dysarthria and Dysphagia.*—Interference with speech and swallowing are prominent features of the disease. The dysarthria is characterized by a slurring of the consonants, and a "cutting off" of the last syllables. Toward the end of the disease there may be a total dysarthria. The dysphagia appears at about the same time that the dysarthria does, and as the condition advances, leads to emaciation and general debility.

In typical cases there are no subjective or objective disturbances of sensation. The reflexes are active, but late in the disease, when contractures have already resulted, cannot be elicited.

MENTAL SYMPTOMS.—These are as characteristic of the disease as the physical symptoms. In some cases they may be insignificant, in others very marked. There is a narrowing of the horizon, docility and childishness. In the chronic type of the disease there is a state of euphoria, associated with restlessness, emotional instability, and a tendency to laughter. In the opinion of Tilney² the changes in the affective

tone and in the expression of the emotions, which are said to be characteristic of the disease and indicative of a lesion in the basal ganglia, do not seem to deserve the importance attached to them, because in 91 cases of pseudo-bulbar palsy published by him, he found in half the cases with no lesions in the lenticular, or caudate nucleus or the optic thalamus, there were typical attacks of laughing and crying, while in one-half the cases with lesions in these parts, no such attacks were observed or reported.

In all of the cases of Wilson's disease there is a marked dementia at the termination of the disease.

The pupils react to light and accommodation. There are no changes in the fundi. The corneæ show pigmentation in a large number of cases. There is, as a rule, no nystagmus and there are no cerebellar symptoms and no sensory changes.

The cirrhosis of the liver is a constant feature, but gives no symptoms during life; this, according to Wilson, is due to the regeneration of the cells in the liver.

Clinical Types.—There are two types of the disease: acute and chronic. The acute is marked by a very short duration, fever and emaciation; it is not as common as the chronic type. The disease is a progressive one, lasting from six months in the acute type to four or five years in the chronic. Relapses and remissions are rare, but they may occur. The prognosis is invariably fatal.

Treatment.—The treatment is entirely symptomatic. Sooner or later the mental symptoms become so marked that the patients must be committed to institutions for the insane.

Pathology.—On autopsy, the liver presents a most striking appearance—a typical "hobnail" liver. The liver involvement is in all cases far advanced; it is strikingly small, nodular and firm. In younger individuals hepatic cirrhosis may sometimes exist as an independent disease and not as a part of Wilson's disease, but then it is easily differentiated by the fact that cirrhosis in children gives symptoms such as jaundice, ascites or gastric hemorrhages, while in Wilson's disease no such symptoms are observed. In all the "Wilson" livers, attempts at regeneration are shown by the active separating of the bile ducts in the connective tissue bands, and also by the mitotic division of the liver cells resembling a formation of irregular masses of cells, in which the architecture of the lobule is lost.

The most characteristic change, however, is the bilateral symmetrical degeneration of the lenticular nucleus, chiefly of the putamen, less of the globus pallidus. The caudate nucleus may be atrophied, but not to the same degree. The external capsule may be involved, but the internal capsule is intact, and the thalamus, save for the loss of the striothalamic fibers and a thinning of the lamina medullaris externa, is normal.

The degree of the degeneration in the nucleus varies in different cases from a discoloration and porosity to marked atrophy or disin-

tegration. In extreme cases, there are a complete degeneration and cavity formation. Microscopically, there is marked proliferation of the glia with degenerative changes, but without evidences of vascular occlusions or inflammatory lesions.

Pathologically, the condition differs from pseudosclerosis in that the degenerated areas of nerve tissue tend to soften. Such is not the case in pseudosclerosis. In the latter there is a sclerosis which is diffuse, the optic thalamus, dentate nucleus, pons and other parts of the central nervous system participating in the disease. In Wilson's disease, the lesion must show no participation of the blood-vessels, and there must be a cirrhosis of the liver.

In the chronic cases the lesions are generally more marked than in the acute cases, but this is not always so. Hunt regards the condition as an encephalitic or gross lesion in the lenticular region, whereas in paralysis agitans, there is a more specialized lesion in the nature of a system disease. In his opinion, paralysis agitans is due to an atrophy of the large motor cells in the corpus striatum, Huntington's chorea to a destruction of the smaller cells, and Wilson's disease is a combination of the two.

Numerous atypical forms of this disease have been reported in recent literature, and there is a great deal of discussion going on amongst neurologists whether the disease, as described by Wilson, should be regarded as a distinct clinical entity (Sachs). Strümpell believes that paralysis agitans, Wilson's disease and pseudosclerosis all belong to one group. Rausch and Schilder, in a number of cases of pseudosclerosis which they studied, found it to be a hereditary, degenerative disease, involving the brain and liver simultaneously. They also believe Wilson's disease to be a well-defined subgroup of pseudosclerosis.

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THE MYOCLONIAS

Symptomatology of clinical types, p. 547—Paramyoclonus multiplex, p. 547—Unverricht's myoclonia, p. 548—Lundborg's myoclonia, p. 549—Myotonoclonia, p. 549—Nystagmus-myoclonia, p. 549—Diagnosis, p. 550—Treatment, p. 550—Prognosis and duration, p. 550—Pathology and pathogenesis, p. 550—References, pp. 551-2.

Definition.—The myoclonias are characterized by the common symptom of rapid, lightning-like, involuntary, clonic contractions of single muscles or groups of muscles, occurring in paroxysms, and which produce no movements or the very slightest movements of the parts affected.

Symptomatology of Clinical Types.—PARAMYOCLONUS MULTIPLEX.—In 1881, Friedreich described a symptom-complex in which lightning-like, jerky contractions of the muscles is a predominating feature, and which he called "paramyoclonus multiplex." The involvement is symmetrical, affecting mainly the muscles of the extremities and the trunk, e.g., the latissimus dorsi, trapezii, gastrocnemii, quadriceps extensors, the pectorals, the recti abdominis, biceps humeri, triceps, etc. The facial muscles, as a rule, are not involved, but the larynx and diaphragm may sometimes be affected. The movements are confined to muscle fibers, bundles, or individual muscles and resemble fibrillary twitchings; at times the contractions cause the bulging of an entire muscle. These twitchings occur at the rate of from 30-100 or more per minute, lasting for several minutes; they may occur every half hour or more often, depending upon the severity of the disease. The jerkings may be arrested by calling the patient's attention to them; they are aggravated by rest and distraction of the patient's attention; they are diminished or may cease entirely during sleep. The oscillations are irregular, vio-

lent ones alternating with weak ones. Although the involvement is symmetrical, the contractions are neither synchronous nor rhythmical. A tap upon a tendon, a touch of the skin or the mere exposure of it will start a jumping and vibrating of the muscles in all directions, and quite independently of one another. The twitchings after such stimulation will appear as if the muscles were shocked by a weak electrical current. No matter how violent the contractions are, they never produce movement in the limbs or joints to which the muscles are attached. L. P. Clarke¹ reports a case of this disease in a patient in whom the myoclonic contractions became most marked and uncontrollable when he turned around in bed to change his position. There are other similar cases reported in the literature.

The reflexes, superficial and deep, the myotatic and electrical irritability are normal; the cranial nerves, and the sensibility are unaffected.

In the cases recorded, males seem to be more frequently affected than females; the symptoms begin at any time of life between puberty and the age of sixty, fifty being the most common age. The disease may occur spontaneously or after fright, trauma (Starr, Carrière, Bertrand), mental or physical exertion and after infectious fevers (Raymond, Valebra, Sterling, Meynert). Many cases are said to occur in Italy after malaria. M. Lafforgue² reports a case in which pronounced myoclonic symptoms appeared in one of his patients on the tenth day after a severe attack of mumps, which was complicated with orchitis. Remak saw a case following diphtheria.

The disease has been met with as a complication of lead poisoning (Leusche), of paresis (Grawitz, Williams), of anterior poliomyelitis (Bailey), of meningo-encephalitis (Clarke), and the writer has seen it complicate Friedreich's ataxia in twin brothers. Lenoble and Aubineau report a case complicated with glycosuria. (Autopsy in this case showed no lesions in the brain or cord.)

UNVERRICHT'S MYOCLONIA.—Unverricht has described a form of myoclonia which is characterized by its familial nature and its association with epilepsy. It has been known to affect successive generations. The twitchings in this special form involve the muscles of the tongue, pharynx and diaphragm. The epileptic seizures occur in the beginning at rare intervals, but become more frequent later in the disease; they occur especially at night. The frequency of the occurrence of the disease may be gathered from the following statistics: Shanahan met the disease 7 times in 2,150 cases of epilepsy, Williams 6 times in 794 cases and Turner twice in 2,000 cases. In many of the cases the paramyoclonus antedates the epilepsy, and in many of the cases the epilepsy escapes recognition, because the seizures occur only during the night; in most of the cases, however, the epileptic seizures precede the myoclonia, which usually develops at the age of ten. As the disease advances, the epilepsy diminishes and the myoclonia becomes more marked. Rhein³ reports a case of familial myoclonus in a sister and

brother in whom the typical myoclonia was not associated with any form of epilepsy.

In Unverricht's myoclonia the twitchings are intensified by the influence of emotion or after physical exertion. The tendon reflexes, myotatic and nerve irritability are increased. Lundborg has emphasized the tendency, which patients afflicted with myoclonus have, to develop a dementia similar to dementia precox late in the disease.

LUNDBORG'S MYOCLONIA.—A variety of myoclonia, which seems to be quite prevalent in Sweden, was studied by Lundborg, who found that, during the early stages of the disease, the patients had alternately good and bad days. He spoke of a "psychoclonic reaction" in which the symptoms are aggravated by emotion, a "psychotonic contraction" during which tonic contractions of certain muscles take place during embarrassment with consequent inhibition of movement, and of a "sensoclonic reaction" in which the myoclonia becomes very intense on the "bad days" after the slightest sensory stimulation.

Popoff⁴ reports a case under the name of MYOTONOCLOIA in which a 19-year-old male had developed, at thirteen, clonic contractions of the left wrist during voluntary movements; later the fingers became involved. Three years after this, tonic contractions developed in the muscles of the forearm, followed by tonic and clonic contractions of the muscles of the neck and finally in the flexor muscles of the trunk; the spasms would last for two months, after which they would disappear to return later. In addition to the myoclonic contractions, there was a tremor involving the entire body, which would increase when the patient attempted to straighten out his body. When sitting, lying down or sleeping the tonic and tremor would both disappear. The psyche was normal and there were no indications of organic nervous disease. The patient's father and younger brother showed similar myoclonic disturbances, but without involvement of the abdominal muscles.

Cases similar to those of Unverricht and Lundborg have been described by Buhrer, Garnier, Verco, Putnam, Mott and others.

NYSTAGMUS-MYOCLONIA.—Lenoble-Aubineau⁵ described a rare and obscure disease which occurs almost exclusively in the Celtic races of Brittany and Great Britain. It is a hereditary familial disease, characterized by spasmodic twitchings of the external muscles of the eye, of the extremities and tremor of the head. Exposure or percussion of the muscles intensifies the twitchings which can, to certain degree, be voluntarily suppressed; the reflexes are exaggerated. Trophic and vasomotor disturbances are not uncommon. Stigmata of degeneration, such as deformed teeth, body and facial asymmetry, local hyperhidrosis, circumscribed edema and lividity of the skin are often present. The disease, although incurable, does not seem to be progressive. No pathological changes have been found in the cases examined.

N. S. Yawger,⁶ of Philadelphia, has recently reported several cases of familial head nystagmus in four generations associated with ocular nystagmus in a Russian-Jewish family. There were several stammerers in the family. Both sexes had the combined nystagmus and both trans-

mitted it to their children. In none of the cases was there spontaneous nystagmus; in most of them the ocular movements, the speed of which was from 120-200, preceded the head movements, of which the patients were not conscious, unless their attention was called to them. The head movements were horizontal, in the same direction as the eye-movements, and coördinated; the approximate frequency being 20 per minute. All the patients were mentally alert and had no other signs of organic nervous disease. Similar cases have been reported by Popper,⁷ Rosenfeld,⁸ Thompson,⁹ and Nettleship.¹⁰

Diagnosis.—The clonic, lightning-like, rapid, symmetrical paroxysmal twitchings of the muscles of the trunk and extremities, without involving the face, and producing no movement of the parts affected are sufficiently characteristic to distinguish the condition from chorea and its allied disorders.

Treatment.—Bromids, chloral and other sedative drugs have a temporary quieting effect on the twitchings. All forms of electricity, particularly the galvanic, have been recommended. Thyroid extract is said to be of service. Hydrotherapy judiciously employed is also of some benefit. Psychotherapy is of service only in cases which are associated with hysteria. The cases which the writer had an opportunity to see were not benefited by any treatment.

Prognosis and Duration.—The prognosis as to recovery is not good. The cases which have been reported as cured were probably hysterical in nature. Remissions and relapses are not uncommon. The disease may last indefinitely; many patients have lived to be 70 years old. Death as a rule is due to deglutition pneumonia or myoclonic spasm of the pharynx (rare involvement). In the older cases, death may be hastened by the onset of a gradual dementia and marasmus.

Pathology and Pathogenesis.—Heilig, on account of the facial involvement in some of the cases at the beginning of the disease, believes it to be a form of hysteria, tic or chorea. Strümpell, Huchard, Fiessinger and others consider the disease identical with hysteria. Hoffman and Böttiger think the condition closely related to or identical with chronic chorea. Farge, Mettler, Williams, Lugaro-Soury and others consider myoclonic contractions a symptom of other diseases. Stadler¹¹ reports two cases in which in addition to the characteristic muscular contractions there was in each case a progressive bilateral atrophy affecting adjacent muscles, not in any particular groups, without fibrillary twitchings, reaction of degeneration or sensory disturbances, and on account of these findings, is inclined to consider the disease similar to myotonia (Thomsen's disease). Popoff also considers it myopathic in origin. Friedreich himself thought it is due to an irritation of the anterior horn cells, and Tutschaninow, who reproduced the twitchings in dogs experimentally by injecting the spinal cord with carbolic acid, is inclined to agree with him.

Homer and Vaulair believe the cause to be a hypersensitiveness of the receptor cells of the spinal cord. Hunt also thinks the disease is spinal in origin. Strasman contradicts this view. Lundborg thinks

the contractions are produced by intestinal intoxication. V. Wagner-Jauregg observed in animals, deprived of their thyroid and parathyroid glands, spasmodic phenomena resembling myoclonia. Raymond thinks that myoclonia may be caused by hysteria, psychasthenia, or epilepsy and may accompany any organic disease of the nervous system. Murri, Seppilli, Patella, Massalongo, Clark and Prout consider the origin of the disease to be an irritation of the motor cortex. F. Schultze suggests that convulsive tic is a monoclonia, while general tic is a polyclonia; he believes Unverricht's myoclonia to be the same as chronic progressive chorea. Dana would regard myokymia a form of myoclonia. Oppenheim considers paramyoclonus multiplex an independent disease, and considers Unverricht's cases as being a special type of it, or a different disease, and he believes that in addition to these there are some obscure diseases in which myoclonic contractions may be symptoms of special importance. He calls attention to the difficulties encountered in the recognition of the hysterical myoclónias.

The results of anatomic examination have thus far simply added to the confusion which exists both as to the pathology as well as to the pathogenesis of the disease. In many cases, intense lymphocytic infiltration was found in the brain and cord. In a case of familial myoclonus epilepsy, Sioli¹² found slight degeneration in the upper cervical cord, the so-called Hellweg triangular tract; the anterior horn cells were normal. The cerebral cortex was like that found in epilepsy, and in the cerebellum he found a large lipid mass near the dentate nucleus, which extended into the white matter of the cerebellum and somewhat into the pons. Sioli does not know whether this lipid mass was a chance finding, or whether it bears some relation to myoclonia. It may also be of interest to note that Hunt found a hypertrophy of the primary fibers in the muscles. In most of the cases, however, no lesions were found, to which the clinical manifestations of the disease could be ascribed.

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THE ATHETOSSES

Etiology, p. 552—Symptomatology, p. 553—Diagnosis, p. 554—Clinical types, p. 554—Treatment, p. 555—Prognosis, p. 556—Pathology, p. 556—Bibliography, p. 556.

Definition.—In 1871 William Hammond, a New York neurologist, described a condition—"characterized by an inability to retain the fingers and toes in a fixed position into which they may be placed, and by their continuous motion, due to involuntary contractions that take place slowly, apparently as if with deliberation and great force"—which he called "athetosis" (*ἀ* = without, *τιθέναι* = to place).

The condition has since then been described and studied by different observers, both clinically and pathologically, but our knowledge as to its etiology and pathology is still very indefinite. It is also known as "mobile spasm."

Etiology.—The exact etiology of the disease is not known. Trauma, chill, infections, physical exertion and mental excitement are said to have preceded some of the acquired cases. A hereditary neuropathic or psychopathic taint has frequently been traced in the family. Oppenheim saw the disease twice in members of one family, and in another family, a mother and daughter were both affected. Massalonga describes three cases in one family. Osler collected 53 cases in the literature, 33 of which were in males and 20 in females. The disease bears no relation to hemiplegia.

It is characterized by spontaneous, bilateral, symmetrical wriggling and twisting movements of the muscles of the face, neck and limbs. The movements are best evinced during attempts at voluntary movements; they are generally accompanied by spastic rigidity of the involved muscles, so that the patients are usually unable to walk and are more or less helpless. The peculiar contortions of the body, with the grimacing of the face, present a striking clinical picture, not unlike that seen in Huntington's chorea. The involvement of the muscles of articulation interfere with speech, and that of the tongue and muscles of mastication interfere with the taking of food. The participation of the diaphragm and of the muscles of respiration results in a peculiar, jerky form of breathing, and the voice assumes an interrupted and groaning character.

Although the disease appears to belong to the group of infantile cerebral diplegias, signs of pyramidal tract involvement are very rare. While the movements may be the only symptom present, a certain degree of mental deficiency, epilepsy or other nervous diseases frequently accompany the condition. There is a strong tendency to associated movements. Lewandowsky (*see Bibliography*) insists that double athetosis is not simply the involvement of both sides of the body with typical athetosis, but that it is a different condition and depends upon excessive associated movements. When undisturbed, the patients may remain perfectly quiet. The condition may manifest itself in the most pronounced form when there is very little motor paralysis.

Symptomatology.—The movements occur most frequently in the spastic extremities of old-standing hemiplegics, and are most commonly observed in children. They are hardly ever seen in a limb which is completely spastic and paralyzed, but only when some degree of voluntary power has survived or returned. The character of the movements has been variously described as "writhing," "cramp-like," "vermicular," "serpentine," "ameboid," "like tentacles of polypi," etc. They are slow, arrhythmical, involuntary twisting spasms, most marked in the fingers and wrist, and in severer cases in the forearm, elbow, shoulder and leg, where the most common movement is an involuntary hyperextension of the great toe. The movements are those of separation and adduction combined with flexion and extension. The fingers are not moved at the same time, nor in the same direction; there may be alternate abduction and opposition of the thumb with flexion or extension of the wrist and pronation or supination of the forearm. The movements rarely involve the muscles of the face and neck, but when they do, they result in the most hideous grimacing and contortions. This involvement is only observed in bilateral athetosis. The constant morbid excess of motion is frequently followed by a hypertrophy of the muscles and a hypermobility of the joints.

Ordinarily the movements are absent during sleep, but in severe cases, however, they may be in evidence even then. During comparative rest they are diminished, and if the hands and feet are interlocked, they may cease altogether. The slightest physical or mental excite-

ment intensifies them; a mere effort to walk will bring out the movements very promptly. Attempts at active or passive movements of the affected and at times even of the unaffected limb, are followed by a temporary interruption of the spasms only to be renewed with greater violence. The patients are sometimes able to restrain, momentarily, the abnormal movements, but they must make a supreme effort of the will to do so; enforced restraint is followed by unusually strong movements.

The leg is always affected to a slighter degree than the arm; the spasm is extensor in type, and the foot tends to assume the position of talipes equinovarus; some fixed rigidity of these muscles frequently coexists. In some cases, instead of the spontaneous spasm, or, associated with it, incoördination of the voluntary movements, varying from a slight ataxia to a jerking, wild incoördination, similar to that of multiple sclerosis may be seen.

Diagnosis.—The diagnosis of the condition, with the characteristic slow, irregular, worm-like movements, involving most commonly the distal ends of a non-completely paralyzed limb, leaving the muscles of the face intact, as a rule presents no difficulties.

Clinical Types.—Two types of athetosis are recognized, the *symptomatic* and the *idiopathic*.

Bilateral, Idiopathic or Primary Athetosis (Athétose double).—This is a rare disease, usually congenital. It may not necessarily appear at birth; it may begin during childhood or even during adolescence, but is most prevalent in infancy.

Under the *symptomatic* type are included the cases that are secondary to a causative factor or a disease. It is much more common than the idiopathic form. It is most commonly seen in the infantile cerebral palsies both of the hemiplegic or diplegic variety. Some of the cases are congenital, being due to developmental defects of the central nervous system, parental syphilis or alcoholism, injuries to the gravid uterus, and difficult labor with injuries during birth. It may be one of the symptoms of a tumor, a gliosis, or a vascular lesion of the brain. It may follow encephalitis after toxic or infectious conditions, especially when these are accompanied with hyperpyrexia. Athetoid movements may be seen in Friedreich's disease and in advanced tabes of the superior type and the bulbar paralytic form. Berger reports an undoubted case following cerebral abscess.

When it occurs in infantile cerebral palsy, it may be so slight as to be merely a suggestion of the condition. The interval between the onset of the paralysis and the appearance of the athetoid movements may be months or years after a certain amount of voluntary motion has returned. Two etiological facts, in the opinion of Gowers, are of great significance. One is, that the movements are seen more frequently after cerebral softening from occlusion of the vessels than after cerebral hemorrhage, and the other, that they follow hemiplegia more frequently in infancy than they do in adult life. The significance of the first fact, he thinks, is that in softening, slight damage to the brain-tissue is

more extensive than the actual destruction, and the movements are due to overaction of the gray matter, which is in a state of altered nutrition and function. The significance of the second fact is probably the greater ease with which the developing brain cells recover, and their greater tendency to disorder of function when their development is perverted.

In the hemorrhagic cases, athetoid movements follow the absorption of the clot and the reestablishment of the circulation, with the consequent scar formation, which gives rise to irritation. When athetoid movements are due to tumors, they are seen in the initial stages of the tumor growth, but disappear later when the pressure exerted by the neoplasm has been sufficient to produce absolute paralysis.

The symptoms depend on the site of the lesion and the nature of the pathological process. Unilateral or hemiathetosis is by far more common than the bilateral variety. The lesion is usually situated in the thalamus of the opposite side. It may be confined to the parts of the internal capsule, immediately adjacent to the thalamus, and also to the lenticular nucleus, rarely in the capsule itself. When, however, the capsule is involved, the lesion is, as a rule, found in the posterior third of its posterior limb. The foci may also be situated in the sensory tracts, which pass into the thalamus, i.e., the cerebello-thalamic tract or the tract of the superior cerebellar peduncle.

These localizations explain the differences in symptomatology, why in some cases the reflexes are exaggerated, and in others diminished or absent, and why some cases have hypertonicity of the muscles with other signs of pyramidal tract involvement and others have not. The presence or absence of sensory disturbances also depends on the anatomical localization. In about half the cases, in which the condition follows hemiplegia, in the adult there is hemianesthesia, or hemihyperesthesia, or hemiparanesthesia, but in the cases which date from childhood, sensation is almost always normal.

The frequently coexisting associated movements, or hemichorea, or hemiataxia are also due to the localization of the morbid foci. At times the movement in the athetotic extremity does not seem to be characteristic of pure athetosis, but there is a phase of an athetoid mixed with a choreiform movement to which the name of "choreo-athetoid" is given. There are also various transitional forms, which are rather difficult to classify.

Treatment.—The treatment is purely symptomatic, and not very satisfactory. **Sedatives, systematic exercises, hydrotherapy, electricity, and ingenious mechanical contrivances** for purposes of immobilization have been employed, but the results have not been of sufficient benefit to recommend their use. **Surgical treatment** has been resorted to by Hammond, Spiller, Media and Bossi, and Sir Horsley, and the good results reported to have followed were in cases in which the movements were limited to groups of muscles or to one limb. In the bilateral type of the disease, in which there appears to be a diffuse involvement of the brain combined with mental deficiency or epilepsy, surgery can promise very little hope.

Prognosis.—The disease is a progressive one; rarely does it remain stationary. All other functions of the body being intact, the prognosis, *quoad vitam*, is good. The disease lasts many years, death being due to some intercurrent affection. Oppenheim has seen a case recover apparently after mercurial treatment.

Pathology.—Anatomic examination has usually given negative results. Oppenheim as well as Déjerine-Sollier found irregularities in the convolutions of the brain. Kurella found in one case a pachymeningitis and a bilateral lesion in the motor convolutions. The postmortem findings must be rigorously differentiated from those of the spastic infantile diplegias that are combined with athetosis. Lesions have been found in the cerebellum and medulla as well as in the cerebral cortex, but no relationship could be established between these and the clinical symptoms. The consensus of opinion seems to be that the disease is due to a bilateral irritation of the motor area.

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MYATONIA CONGENITA (OF OPPENHEIM)*

History and definition, p. 556—Occurrence, p. 556—Symptomatology, p. 556—Diagnosis, p. 557—Treatment, p. 558—Course and prognosis, p. 558—Pathology and pathogenesis, p. 558—Bibliography, p. 559.

Synonym.—Amyotonia congenita.

History and Definition.—In 1900, Oppenheim drew attention to a condition characterized by marked muscular atony and a peculiar paralysis occurring in early childhood, and closely resembling the clinical picture of poliomyelitis.

Occurrence.—The disease is noticeable at birth, but typical cases have been known to have begun in the first and second years of life.

Symptomatology.—The involvement is most marked in the muscles of the lower, sometimes also of the upper extremities, and less often in those of the neck and trunk. Involvement of the head, face, tongue,

*This is not to be confused with myotonia congenita (Thomsen's disease) or with dystonia musculorum deformans, the three diseases being distinct conditions.

eyes and larynx have not been observed. The muscles do not appear atrophied, but their flaccidity is very striking. The atony is so marked that the extremities can be placed in the most bizarre positions, as if they were loosely-attached appendages. When the little patient is seated the trunk bends forward, forming a marked kyphosis. That the paralysis is not a true one, but rather a weakness, can be demonstrated by placing the child on his feet, when it will be seen that not only is he unable to stand alone, but even when supported, his legs give way under him; but when he is placed on his back, feeble active movements of the arms and legs can be carried out. The hypotonia of the various joints is also a very striking symptom.

The cranial nerves, the sphincters and sensation are not involved; there are no fibrillary twitchings. The deep reflexes are absent, while the superficial ones remain normal. The direct and indirect electrical excitability, both to faradism and galvanism in the paralyzed as well as the apparently sound muscles, is usually reduced and frequently absent; there are no polar changes. In one of Oppenheim's cases, while the legs lay as if paralyzed, previously absent knee-jerks could, after electrical stimulation, be freely brought out.

Purser¹ met a case of this disease in a two-year-old child, in which the condition developed at fifteen months, after an attack of severe diarrhea, lasting four weeks. The patient had rickets and an unusual symptom—nystagmus. According to Purser, this was the first case of myatonia reported up to 1914 in Ireland.

Diagnosis.—Collier and Gordon Holmes (cited below) point out the following clinical features of the disease which distinguish it from the *myopathies*: (1) The absence of any familial tendency. (2) The disease is in the majority of cases congenital, and in a small number of cases it appears suddenly and is fully developed after certain acute diseases. (3) The local wasting and weakness of an individual muscle or of a group of muscles that are characteristic of all forms of myopathy are not met with in myatonia. (4) Affection of the periphery of the limbs, and especially of the intrinsic hand muscles, which is the invariable rule in myatonia, is of the greatest rarity in any form of myopathy. (5) Myatonia never spreads to regions previously unaffected, slow spreading of the affection from muscle to muscle being characteristic of all forms of myopathy. (6) The deep reflexes are absent from the beginning, but may reappear after improvement in myatonia, while in the myopathies they are present at first and slowly diminish as the condition progresses, and finally are lost, never to be regained. (7) There is a tendency to improve in myatonia, and in some cases recovery may ensue; this is never the case in myopathy.

The absence of atrophies or sensory changes, the generalized symmetrical involvement, and the electrical changes which can be seen in mostly *all* the muscles distinguish myatonia from *anterior poliomyelitis*, and from the peripherally and spinally induced *intrapartum paralyses*.

Myatonia must also be distinguished from *syphilitic pseudoparalysis* as described by Vierordt, and *muscular weakness due to rickets*, as well

as from joint hypotonia due to involvement of the capsules and ligaments of the joints (Finkelstein).

Treatment.—General hygienic and tonic measures are indicated. Careful medication with **strychnin, massage and electricity** are resorted to. **Orthopedic appliances** may be necessary. Powis and Raper found, after the administration of **bile salts or dried ox bile**, an increase in muscle strength.

Course and Prognosis.—The course of the disease is slow; the prognosis as to life is good. In some of the cases, in the course of time a more or less complete recovery may occur.

Pathology and Pathogenesis.—Spiller,³ in his first autopsy, found changes in the muscles only, and none in the nervous system. Baudouin⁴ found nothing characteristic for pathologic inferences. In another case Spiller and Griffith⁴ found atrophy of the muscle fibers much more marked in some bundles than in others. The atrophied fibers retained their transverse striations and showed an excessive number of nuclei in the sarcolemma; some of the muscle fibers presented fatty degeneration. These were the changes found in the muscles of the forearm; the atrophy was most marked in the calf muscles, and least marked in the muscles of the back. Accompanying these muscle changes was a small spinal cord, with scanty and atrophied anterior horn cells; the anterior roots were smaller than ordinarily and stained very poorly. One posterior tibial nerve was examined and also found much degenerated. The brain in this case was unusually large. Skoog, Reyer and Helmholtz found similar changes in the muscles "*in vivo*," whereas Bing found the muscles to be perfectly normal.

Collier and Gordon Holmes⁵ in a study of 2 cases found most of the muscle fibers smaller, which they say might be attributed, on the one hand, to a lack of development, or, on the other hand, to atrophic processes affecting them; but they are more inclined to believe, on account of the extraordinary irregularity in the shape and size of the fibers, that the condition was due to an atrophy. The atrophy and decrease in the number of fibers, in their opinion, sufficiently explains the palsy, while the fatty infiltration and the increase of connective tissue cells conceals in part the general atrophy.

De Villers⁶ had 2 cases, which confirmed Concetti's conclusions, based on 68 cases with 16 autopsies, that the cause of the disease is an arrest of development or retardation during intra-uterine life of the anterior horn cells of the spinal cord, and that the lesions may invade the cerebrum, cerebellum, peripheral nerves and muscles, but that there is a tendency to progressive improvement.

Oppenheim believes the disease to be due to an arrest of development of the muscle fibers. Marburg held that the cases were forms of intra-uterine poliomyelitis. The symmetrical development of the defects in all cases, the development of some of the cases after birth, and the fact that in a good many instances the anterior horn cells were not involved, and in those cases in which they were involved there was

no evidence of inflammatory changes, speak against the correctness of Marburg's theory.

Some investigators are inclined to ascribe the disease to a disturbance of the ductless glands. Berti considers it a variety of congenital myxedema. In this connection it may be of interest to note that Powis and Raper⁷ were led to the following conclusions from their metabolic studies in this disease: (1) There is a diminution in hepatic function, as evidenced by the presence of acholia; (2) normal calcium retention is associated with a relatively high potassium retention; (3) there is a low creatinin excretion with a relatively high creatin excretion.

Rothman believes the cases to be a type of Werdnig-Hoffman's disease; the fact that there is no wasting of the muscles and that there is a tendency to improve, after a time, speak against this contention. A. Gordon⁸ believes that there is an antenatal disturbance of nutrition of the anterior horn cells, which in some cases leads to their destruction, thus producing a Werdnig-Hoffman type; in other cases the nutritional disorder is slight, resulting in a diminution of muscle tonus. He also believes that there are a large number of intermediary cases difficult to classify clinically.

Bernhardt is inclined to assume an injury to the peripheral nerves, somewhat like a generalized polyneuritis of autotoxic or infectious origin.

From the facts adduced it is evident that the pathology of the disease is not definitely known, and the same may be said of its causation. There is a tendency to consider the condition a type of myopathy, rather than a distinct clinical entity.

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DYSTONIA MUSCULORUM DEFORMANS

Occurrence, p. 560—Symptomatology, p. 560—Treatment, p. 561—Prognosis, p. 561—Pathology and pathogenesis, p. 561—Historical summary, p. 562—Bibliography, p. 562.

Synonyms.—The disease has been described by different observers under different names, such as *Dysbasia lordotica progressiva*, *Dystonia musculorum deformans* (Oppenheim), *Tonic torsion spasm* (Ziehen), *Progressive torsion spasm of children* (Flatau-Sterling), (Hunt), *Ziehen-Oppenheim disease* (Van Bernstein), and *Tortipelvis* (Fraenkel).

Definition.—A symptom-complex characterized by disturbances of muscle tone, resulting in bizarre movements of groups of muscles.

Occurrence.—The disease affects mostly children of Russian-Jewish parentage between the ages of eight and fourteen. Diller and Wright, Patrick, Dereum and others report cases in which the disease occurred between the ages of twenty and thirty, but its occurrence at this age is rare. Of the number reported hitherto, it seemed to have predominated in females.

Symptomatology.—The affection is characterized by a *deformity* around the pelvic girdle, with tonic and clonic spasms, involving chiefly the flexors of the thigh, pelvis and lumbar region. Other muscles may be affected, but not as frequently as the muscles concerned in locomotion. The affected muscles are alternately in a state of hypertonia and hypotonia. On standing and walking, the so-called “dromedary” or “monkey” gait presents a striking picture. The disease may occur in the form of a monoplegia, diplegia or hemiplegia. The twitchings usually begin in one upper extremity, and gradually other muscles become affected, but the involvement is most marked in the muscles of the pelvic girdle. The muscles of the face, of speech, mastication and swallowing usually escape. The author has reported a case in which, in addition to an extensive quadruplegia, all the muscles of the face, neck, tongue and swallowing participated in the disease (*see Bibliography*). The twitchings are evident while standing and are intensified on voluntary movements and mental excitement. The constant spasm and the deformities which result from them, such as scoliosis, lordosis, tilted pelvis, etc., are diminished during rest, and cease entirely during sleep.

The twitchings may be in the form of a rhythmical tremor or rhythmical clonic contractions. The fatigue and strain, brought on by the spasms, increase the rate of the pulse and produce vasomotor disturbances, such as flushing of the skin and hyperidrosis.

There are no signs of organic involvement of the nervous system: the tendon reflexes are normal, but at times difficult to elicit, owing

to contractures which may exist. There is no paralysis, no muscular atrophy, and no electrical changes. The sensations, sphincters and cranial nerves are all intact. There are no mental disturbances.

All volitional movements are performed as if there were a conflict in action of the different muscle groups, a reversal of muscle tonus. There is an inability to dissociate harmoniously the essential muscle tonus elements of a simple movement, namely, contraction of the agonists and antagonists. This Hunt designates as the "paradoxical" or reverse phenomenon of dystonia, and he considers it diagnostic in the differentiation of this disease from spastic, hysterical and other forms of spasmodic contractures. This sign is elicited in the following manner: The wrist joint being held in flexion by the patient when he is requested to extend the hand or the forearm, he makes an attempt to do so, but instead of extension there are several involuntary flexions to a greater angle than there was originally, and after a perceptible lapse of time the desired extension is first carried out. The same result may be obtained in any other joint.

Treatment.—Therapy has had little or no effect. Psychotherapy, electricity, hydrotherapy and metallotherapy have been employed, but only with temporary benefit. In one of Fraenkel's cases, reëducation seemed to help for a time, as did the intraspinal injection of magnesium sulphate. Sedatives, such as bromids, hyoscin and even morphia, are occasionally indicated, to give these patients relief from the severe spasms. Luminal in large doses has been administered by the writer, but with no effect. In one of the cases at the Montefiore Home and Hospital the spasms were so severe that it seemed as though the patient would die of asphyxiation at any moment. **General anesthesia with chloroform** has been resorted to; relaxation occurred after the patient had been completely anesthetized. The patient stood the anesthesia very well, but upon its withdrawal and before full consciousness had been regained, the spasms recurred. In one case the application of a plaster of Paris cast to prevent contractures aggravated the condition. In all cases it seems that mental and physical excitement have a tendency to bring on an exacerbation of the disease.

Prognosis.—The prognosis, *quoad vitam*, is good. The patients, as a rule, die of some intercurrent disease. The disease is chronic, progressive, and incurable. There are cases in the literature in which improvement occurred spontaneously or after treatment, but such improvement was only temporary, to be followed by remissions, which were just as severe as the original attack.

Pathology and Pathogenesis.—There is nothing definitely known of its etiology or pathology. Oppenheim believes the disease to be due to fine pathological changes in the cells of the cortex which control muscle tone. Fraenkel thought the disease to be due to faulty calcium metabolism. Studies of the calcium metabolism undertaken by Janney at the Montefiore Home and Hospital, on one of the patients in that institution afflicted with a severe type of this disease, showed no abnormality in the elimination of this mineral.

Dana believes the disease to be a neurosis allied to general tic. Seclert thinks the disease is psychogenic in nature. Although the disease appears to Bonhoeffer at first sight to be a form of hysteria, prolonged observation leads him to class it among the choreas. Bing considers the condition to be similar to athetosis.

Jelliffe thinks that the lesion is in some portion of the cerebello-thalamo-cortical arc, probably cortical to the red nucleus and possibly in the region of II' and II'' of Forel's field.

The disease is included by many as one of a number of diseases due to a disturbance of tone and associated as such with the "amyostatic" syndrome of Strümpell.

J. Ramsay Hunt, after an exhaustive study of six cases of the disease and a thorough review of the literature, says that he is convinced that "the lower type of mechanism is at fault, one which is closely associated with the corpus striatum, but which is engaged in the regulation of tonus, especially in its relation to the reciprocal activities of the agonistic and antagonistic muscles, for this constitutes one of the essential factors in the motor disturbances of torsion spasm; but the preservation of facial expression and articulation is against a localization in the corpus striatum, for we know from many pathological studies that the face and articulation are both involved in this localization."

Up to the present time there is only one record of an autopsy and that was in one of Ziehen's cases. No lesions of any kind could be demonstrated in the brain or cord.

Historical Summary.—At a meeting of the Berlin Psychiatric Society, held in December, 1910, Ziehen reported 5 cases of peculiar twisting spasms in children, which he called "torsion spasm." Three of these cases had been previously published by Von Schwalbe in 1908. In October, 1911, Oppenheim published the clinical histories of four similar cases, and outlined a symptom-complex to which he gave the name "Dystonia musculorum deformans." *The disease has since then been called Oppenheim's disease. This name is unfortunate because this symptom-complex of dystonia is apt to be confused with that of myatonia congenita, which is also sometimes described in the literature as "Oppenheim's myatonia" (see chapter on Myatonia).* In his original report Oppenheim stated that he had seen similar cases before that time, but that he did not know whether they were cases of hysteria, hysterical scoliosis, lordosis or idiopathic bilateral athetosis. Since then, numerous cases belonging to this group have been reported both abroad and in this country.

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MYOTONIA CONGENITA (THOMSEN'S DISEASE)

Etiology, p. 563—Symptomatology, p. 564—Myotatic irritability, p. 565—Electrical reactions, p. 565—Diagnosis, p. 565—Varieties and clinical types, p. 566—Myotonia acquisita, p. 566—Pseudo-myotonia hemiplegica, p. 566—Myotonia atrophica, p. 566—Paramyotonia congenita, p. 568—Treatment, p. 568—Prognosis, p. 568—Pathology and pathogenesis, p. 568—Historical summary, p. 569—Distribution, p. 569—References, p. 570.

Synonyms.—Thomsen's disease, Myotonia atrophica.

Definition.—Myotonia congenita is 'a hereditary disease characterized by muscular cramps at the beginning of voluntary movements, which disappear on repetition of the movements.

Etiology.—PREDISPOSING CAUSES.—*Sex.*—The disease has a preference for the male, beginning in typical cases usually in early youth or about the time of puberty.

Mental Shock.—The disease is said to develop after mental excitement or physical overstrain; this is probably due to the fact that the shock or fatigue has increased the disease which had already been in existence, but the symptoms were slight, and they escaped detection. Patients afflicted with this disease are generally unusually well developed, full-blooded and well nourished; their muscles are unusually large, but their power is markedly diminished.

Hereditary Influences.—The malady is hereditary, and usually affects several members of the same family. Because the disease is not

always congenital, Gowers suggests that the name "transient myotonia" would be a more appropriate one. Blood relationship in the parents has, in certain cases, been the cause of a marked predisposition, while in other cases no such predisposition could be established.

Symptomatology.—The first and most essential symptom is *stiffness in the muscles*. If after a prolonged rest the patient attempts to carry out a voluntary movement, the muscle or group of muscles called into activity for the purpose of such a movement assumes a condition of *tonic* contraction, which cannot be voluntarily relaxed. After a period varying from 5 to 30 or more seconds, the spasm begins to yield, and the movement becomes easier and "smoother" with each repetition of the attempt, until finally it is carried out without any difficulty. This peculiar contraction, spasm or rigidity is called "myotonus," and is at its height during the *second* and not the first movement. For instance, if the patient wishes to carry out a quick and strong movement with the thumb, he may succeed fairly well with the first adduction of the thumb, but the succeeding abduction will meet with very strong resistance, which can only be overcome upon the further repetition of the movement, which then becomes freer and freer until the tension is completely relaxed and the movement is eventually carried out in a perfectly normal manner.

The myotonus is very marked when the patient is suddenly called upon to make a rapid and forceful complex act, such as attempting to rise and walk after he has been sitting in a chair for a time. Going up and down stairs, with the alternate relaxation and contraction of the muscles of the leg, is very difficult. These patients must be very careful on crossing streets busy with vehicular traffic or on getting on and off cars; in the latter case it may be impossible for them to release the hand rail with sufficient rapidity to avoid being thrown against the car when getting on, or to the street when getting off.

Excitement, fright, dampness and cold aggravate the myotonus; mental rest, warmth and the ingestion of small amounts of alcohol diminish it. The condition is worse when the patient thinks he is being observed. The more prolonged the rest, the severer the spasm; when it has once passed off the patient can carry out the most complicated and most delicate acts without the slightest difficulty or fatigue.

The affection may involve all the muscles of the body, but it is most frequent in the limbs, and least in the muscles of the face and jaw. The ciliary muscles may be involved, so that the patients find it difficult to accommodate for objects at different distances. In one of Charcot's patients, the eyeballs, on looking upward, remained in that position for a considerable length of time. Involvement of the tongue is not uncommon, but rarely are the muscles of the throat and respiration affected. The heart muscle is said to have been involved in some cases: **Boot**¹ reports a case of this disease in a boy in whom the heart sounds were unusually loud. The arms may be free and the legs affected, or *vice versa*. There are cases in which the disease is limited to certain groups of muscles. Oppenheim, Gaupp, Schott, Curschmann and others

have described such atypical cases of partial myotonia, but most of them were associated with muscular atrophies. Sometimes the spasm is more marked on one side of the body than on the other. According to Thomsen, the more the muscles are used the less severe is the spasm.

The *myotatic irritability* of the muscles is markedly increased. Percussion of a muscle promptly gives rise to a slow but persistent tonic contraction; the belly of the muscle protrudes like a tumor, or a deep depression or furrow results where it has been struck. These phenomena are most noticeable in the tongue, the thenar and hypothenar eminences and the gastrocnemii. The slightest mechanical stimulation will have the same effect; the muscular contractions may last from 5 to 30 seconds, instead of relaxing promptly, as in health.

Mechanical or electrical stimulation of the nerve trunks elicits no peculiar changes in their excitability, so that no myotonus follows *indirect* electrical stimulation. The change in muscle irritability following *direct* electrical stimulation is a prominent and most characteristic symptom of the disease. This is known as Erb's myotonic reaction "MyR."

The electrical excitability of the muscles is markedly increased for both currents. To galvanism $KCC = ACC$ (in health $KCC > ACC$); the contractions are *sluggish* and continue long after the stimulus has ceased. The stable galvanic current produces a rhythmic undulation of the muscle and the wave of contraction passes from the cathode to the anode; strong currents are necessary to produce this phenomenon. With the usual faradic (direct) stimulation, employing strong currents, there is a somewhat slow contraction with a long prolongation (2 to 20 seconds or more); single opening shocks of the slightest strength cause only normal quick contractions; with strong, stable faradization, oscillating muscular waves are occasionally seen. There is practically no change of reaction to static electricity.

There are no symptoms referable to the nervous system. The cranial nerves, sensation and sphincters are unaffected; the superficial reflexes remain unaltered; the tendon reflexes are active, but they have been reported diminished or easily exhausted. There are no trophic and no vasomotor disturbances.

Patients with this disease are said to take anesthetics very badly.

There are no psychical disturbances. Although Thomsen himself classified the disease under the psychoses, it seems that when psychical disturbances are present they are to be considered complications rather than symptoms of the disease. The condition, while not painful, is a source of great annoyance and, as Thomsen says, "it casts a shadow over the lives of the sufferers."

Diagnosis.—When the clinical features of the disease with the characteristic electrical changes in the muscles are taken into consideration, the diagnosis presents no difficulties. E. W. Taylor² records the case of an Italian shoemaker, 24 years of age, in whom the disease was considered for a long period to be hysteria, because the spasms occurred in paroxysms, between which the patient showed no signs of myotonia.

and because there were no other members of the family afflicted with a similar condition.

Varieties and Clinical Types.—Numerous symptom-complexes resembling myotonia, which are difficult to classify, are described in the literature. There are many conditions, such as tetany, tabes (Curschmann), muscular atrophies, syringomyelia, multiple neuritis, etc., in which a myotonic reaction may be obtained in some of the muscles; this naturally raises the question, whether in these instances we are dealing with complications of myotonia or with different but distinct clinical types. The mode and time of onset of the symptoms, as well as the course of the disease, have led to the description of different types of it. Jacoby, for instance, makes a sharp distinction between the congenital, acquired and transitory forms. He reports a case which developed after an attack of typhoid fever, and another case which followed injury and overstrain. Beco³ saw a case develop in a young man with no history of the disease in his family one year after he had suffered from two abscesses in his foot.

MYOTONIA ACQUISITA.—O. Ascenzi⁴ believes Thomsen's disease as a name should be applied only to congenital myotonia, and this should be differentiated from acquired myotonic states associated with other affections. He found, up to 1912, thirty-one cases, besides one of his own, associated with muscular dystrophy. Hoffman, Rossolimo and Schott think that progressive muscular atrophy may itself develop from myotonia. Talma, also, describes an acquired form of this disease, myotonia acquisita, in connection with traumatism and acute infections, in which the rigidity persisted even during rest. The condition was transient and curable.

PSEUDO-MYOTONIA HEMIPLEGICA.—Quensel⁵ records a case which he calls pseudomyotonia hemiplegica in a man of 45 years who had a peduncular hemorrhage, with disability on the left side, and in whom myotonic contractions always followed voluntary movements. Der-cum⁶ reports a case in a boy of 9 years without a family history of myotonia, which followed an attack of parotitis, with pain and swelling in the knee; there was a typical myotonic reaction with involvement of the arms and legs, but without atrophies or hypertrophies.

MYOTONIA ATROPHICA (Amyotrophic Myotonia).—One of these types known as *myotonia atrophica* was described by Rossolimo⁷ for the first time in 1902 as a distinct clinical entity. Although this type is not as common as the congenital myotonia, it has certain peculiarities which merit a more or less detailed description of it.

Definition.—Myotonia atrophica is a disease, intermediate in its character between the myotonias and the myopathies, in which there is increased tonus in some muscles with a primary flaccid palsy in others, in a more or less constant distribution. Its frequent combination with cataracts is also a characteristic feature.

Frequency.—Batten and Gibb⁸ were able to enumerate 20 cases of the disease in the literature, up to 1909. In this country it is very rare. J. R. Hunt,⁹ of New York, reported 2 cases. Kennedy-Ober-

dorf¹⁰ reported 2 cases, and Kennedy¹¹ himself reported an additional case.

Symptomatology.—Greenfield¹² was the first one to call attention to the frequent association of familial and hereditary cataracts with myotonia atrophica. He reported a remarkable family of 13 members, of whom 6 were healthy, 2 had premature cataract, 3 myotonia atrophica, and 2 had both conditions. Tetzner, Ormond, Hirschfield, Bramwell, Addis and Fearnside described cases in which this association was a marked feature. Hoffman, in 1912, analyzed all the cases of myotonia recorded up to that date, and found that this combination with cataract occurred in 8 out of 80 cases. This association is too frequent to be considered a mere coincidence; it is rather suggestive that both myotonia and cataract may be abiotrophic in character. Another example of abiotrophy is found in a case reported by Bullowa, of New York, in which the disease was associated with a maldevelopment of both testes.

In contrast to the congenital form, the atrophic form does not appear until the second or third decade of life. Grund saw a case appear at 47. Males are more often affected than females. The myotonia generally precedes the atrophies, and as the disease progresses it is found that there are simultaneous atrophies in some muscles with myotonic disturbances in others. The atrophies are most commonly seen in the orbicularis palpebrarum and oris, the temporals, the masseters, the sternocleidomastoids, the pectorals (Abrahamson), the vasti of the thighs and the anterior tibial extensors. In some cases the atrophy may assume the typical type of Erb's dystrophy, or it may be of the Aran-Duchenne type. Griffith¹³ saw a case of myotonia atrophica in which the myotonia was accompanied with an atrophy in some muscles and a marked hypertrophy in others; he called this case "myotonia atrophica and hypertrophica."

Kennedy¹⁴ pointed out a peculiar feature of this disease in that there was an extraordinary uniformity in the appearance of the patients; they all looked as if they belonged to the same family—like brothers and sisters. There are two typical cases of the disease in the neurological wards of the Montefiore Home and Hospital at this writing; they were both shown before the New York Neurological Society at one of its meetings, by Doctor Abrahamson,¹⁵ and the reader is referred to this report for a detailed description of the disease.

The myotonic electrical reaction is modified in that the wasted muscles show the usual diminished irritability to the faradic current and to a lesser degree to galvanic stimulation; occasionally a myasthenic reaction may be demonstrated. Some of the cases show, in localized muscle groups, a modified myotonic electrical reaction, namely, a slow contraction and a slow relaxation to both forms of electrical stimulation. According to Curschmann, some cases show an electrical overexcitability in the nerves.

The condition of the reflexes depends upon the degree of wasting in the atrophied muscles. There are no sensory changes, and no symp-

toms referable to the cranial nerves. The sphincters are not involved, and there are no mental symptoms.

Pathology.—Pathologically it is similar to the myopathies, as far as the muscles are concerned. Steinert found also degeneration of the posterior columns of the cord. There is nothing definitely known of its etiology or pathogenesis. As in all diseases of obscure origin, the glands of internal secretions have been held responsible for the causation of the disease; one such case was studied with this aspect in view at the Montefiore Home and Hospital, but the findings did not differ materially from those in the other forms of myopathy.

PARAMYOTONIA CONGENITA.—In 1886 Eulenberg¹⁶ described a hereditary-familial affection known as “paramyotonia congenita,” which is characterized by a tonic spasm appearing in certain voluntary muscles of the face, especially the orbicularis palpebrarum and oris, so that the patients are unable to open their eyes or to speak for a quarter of an hour or longer. The spasm usually comes on in cold weather, and is not influenced by excessive exertion; warmth has a tendency to diminish it. The muscles of the neck, of deglutition, as well as of the limbs, may occasionally be involved.

The mechanical irritability of the muscles is not increased; but electrical examination shows a diminished excitability. The pathological findings are somewhat similar to those of myotonia congenita. The characteristic symptoms of the disease make their appearance at birth. The disease has been found coincidentally with Thomsen’s disease in members of the same family. (See Delprat¹⁷ and Solder¹⁸ for further references.)

Treatment of the Myotonias in General.—Gymnastics and massage have been employed by Oppenheim and Bechterew. Frink reports the administration of **thymus extract** in one case with good results. Gessler proposed, in the case with severe spasms, to bring about an atrophy of the muscles by **stretching the nerves**; his proposal has been rejected. Johnson and Marshall suggest the use of **strychnin**. In one of the atrophic cases under the writer’s care at the Montefiore Home and Hospital, all forms of treatment—hydrotherapeutic, gymnastic, electrical, gland extracts, strychnin, etc.—have been employed without the slightest sign of improvement.

Prognosis of the Myotonias in General.—The prognosis as to life is good. The disease is chronic and progressive; remissions are not uncommon. The cases which have been reported as improved were undoubtedly atypical forms following trauma or infections.

Pathology and Pathogenesis.—The symptoms of myotonia have been reproduced in animals after poisoning them with veratrin and creatinin. Many investigators believe the condition to be due to an exaggerated excitability of the sarcoplasm. Erb and others have found an enormous hypertrophy of all the muscle fibers; in some cases these were twice the normal size. They also found a profuse proliferation of the nuclei in the sarcolemma, indistinct striation, vacuolization and a slight increase in the connective tissue. Some investigators found atrophic as

well as hypertrophic fibers. Schiefferdecker believes that the increase of the nuclei is only a relative one, but he was able to demonstrate in the sarcoplasm granules which are not seen in normal muscle tissue.

Jacoby also found muscle changes, but is of the opinion that the disease is due to an embryonal maldevelopment of the nerve cells, diminishing their resistance to certain toxic processes in individuals predisposed to the disease. Bechterew is inclined to trace the disease to auto-intoxication.

Metabolic studies have not shed much light on the pathogenesis, except that the empirical administration of thymus extract has been followed by a diminution of the spasms.

Curschmann is opposed to the theory of the myogenic nature of the affection, and he is inclined to ascribe it to a supranuclear or central origin. Although Johnson and Marshall¹⁹ found distinct muscle changes, they are nevertheless of the opinion that the disease is most probably due to a partial obstruction in the higher motor paths. Findlay²⁰ believes that the disease is primarily one of the muscles, but that the nervous system plays no insignificant part in its causation. His reasons for his conclusions are: (1) The creatin and creatinin metabolism shows that we are not dealing with a degenerated but with an unusually efficient muscle. (2) During voluntary movements of the abdominal muscles, such as bending, the myotonic reaction develops very promptly but during involuntary acts, such as defecation, no myotonic contraction of the muscles is observed. (3) The condition gradually disappears on the repetition of voluntary movements, but does not do so on the repetition of a direct or electrical stimulus. (4) The influence which the mental state of the patient has on the condition.

No changes have ever been found in the central or peripheral nervous system, and although the disease is usually placed among the affections of the muscles, its true nature is still to be determined.

Historical Summary.—The disease was first described by Charles Bell and Leyden, but was actually "discovered" in 1876 by the Silesian physician, Thomsen, who was afflicted with the disease himself, and in whose family he could trace it to 20 cases in five generations. To Erb, however, belongs the credit of having classically worked up the disease, especially as regards the electrical changes in it.

Distribution.—Up to 1914, H. Koch collected more than one hundred cases. The disease has been met with in Germany, Austria, Italy, France, Russia, Sweden, England and America. The original reports seemed to have been more numerous in Germany and in the Scandinavian countries than in other localities. The disease is considered to be a rare one, but this is probably due to the fact that some of the cases are so slight that they do not attract attention until perhaps later in life, especially in military countries, when the male citizens are called upon to perform military service.

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SPASMS

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LOCALIZED MUSCULAR SPASMS

General Considerations.—In general, it may be said that a "localized spasm" is a symptom and not a disease. It is defined by Brissaud as a reflex movement due to an irritation along the pathway of a peripheral reflex arc. The movements of a spasm are brusque, resembling the muscular contractions following electrical stimulation. The muscles involved correspond to the anatomic distribution of a nerve. If a portion of one nerve is involved, the spasm may be partial or fascicular, as in spasm complicating facial palsy; if the whole nerve is involved, the convulsive movement may be similar to the performance of a purposive act. In facial spasm, the forehead of the affected side is wrinkled by the contraction of the frontalis muscle, while the eye is closed by the contraction of the orbicularis palpebrarum, both of these muscles being innervated by the same nerve—the facial. Such combined movements cannot be produced at the same time voluntarily. This phenomenon is called by Babinski "paradoxical synergia."

Facial Spasm.—SYNONYMS.—*Spasmus facialis*, Convulsive tic.

ETIOLOGY AND SYMPTOMATOLOGY.—Facial spasm may be the result of organic disease in the area of distribution of the sensory branch of the trigeminus, such as the cornea, conjunctiva, the nose, the teeth, the maxillary bones, the tonsils, etc. It is therefore often associated with "tic douloureux." Any original physiological reflex movement in this distribution may gradually develop into a spasm involving the facial muscles.

Facial spasm begins with clonic contractions, which, as they advance, gain in rapidity, and at the height of the attack become tonic; as this subsides, the clonic movements reappear and remain until the attack is over; the entire cycle lasting about a minute. The twitchings may be observed on the return of power in previously paralyzed muscles, and they may be sufficiently severe to involve the muscles of the other side of the face. Partial facial spasm may be due to overstraining of individual muscles, such as the orbicularis palpebrarum in the course of certain occupations, commonly seen in watchmakers, microscopists, etc.; the spasms are then considered as symptoms of an "occupation-neurosis."

A reflex facial spasm is sometimes noticed in diseases of the ear; it may follow a direct injury to a peripheral nerve, or injuries to the head, which have given rise to organic disease of the brain, or to a traumatic neurosis, i.e., merely from the shock of the injury. A good example of a facial spasm following irritation of a nerve trunk is found in recent literature in two cases of convulsive facial spasm reported by Cushing,¹ due to tumor of the cerebellopontine angle.

Affections in other parts of the body, such as disease of the generative organs, pregnancy, etc., may give rise to facial spasm. It may also be one of the complications of chorea, epilepsy, hysteria, migraine, the psychoses, etc. In many cases no pathological basis for the condition can be discovered, and it is then considered psychogenic; it has been known to develop after shock or great emotional excitement in neuropathic individuals, although direct inheritance is not common.

The most frequent form is hemispasm, confined to the orbicularis palpebrarum, although it may be bilateral, or diffuse and extend to the platysma, occipital and ear muscles. When it is limited to the orbicularis palpebrarum, and when the contractions are purely tonic in nature, it is called "blepharospasm," but when clonic, it is called "blepharoclonus" or "nictitation."

Mental excitement and physical overstrain aggravate the condition; distraction of the attention and suggestion temporarily inhibit it; it may persist during sleep. A sudden examination of the eyes, or a tapping or stroking of the face will provoke it very promptly. Spontaneous remissions and exacerbations are common.

The *subjective symptoms* may be slight or severe, depending upon the intensity of the spasm; true facial spasm may be associated with or provoked by pain in the trigeminal area. There is, as a rule, no

interference with the voluntary movements of the implicated muscles. There are no vasomotor and no trophic disturbances.

TREATMENT.—If a **cause** can be found, it is to be **removed** whenever possible. In general, careful **dieting**, **change of climate**, **change of environment**, with a minimum amount of mental and physical work, and well regulated **hydrotherapeutic** measures are indicated. **Arsenic**, **bromids** and the **analgesics** may be resorted to. **Local subcutaneous injections of cocain** and **atropin** have been used with success as far as the spasm was concerned but the injections have been followed by paralysis of the local parts.

Within the last decade, **injections of alcohol** (70-80 per cent.) into the sheath of the facial nerve on the involved side have been used with success by various clinicians. A most careful technic is necessary. The method was originated by Schloesser;² a few drops of the alcohol is injected along the stylomastoid process as far as the base of the skull, slowly and continuously, until paralysis sets in; this paralysis of the muscles passes off gradually and the patient is relieved for from three to seven months, when the injection must be repeated.

In some of the more obstinate cases, when the supraorbital nerve has been found to be tender on pressure, and when during such pressure the spasm is relieved, resection of that nerve has been successful; simple section of the nerve, however, has no effect. **Stretching of the facial nerve** has resulted in some cases in a temporary improvement; the spasm, however, was followed by a permanent paralysis. Oppenheim prefers injections of alcohol to nerve stretching.

In recent cases, **counterirritation with plasters** behind the ear, the **galvanic current**, **diathermia**, **faradism**, **static electricity**, **D'Arsonval current**, etc., have been recommended.

Grossman, of New York City, has reported a large series of cases in which improvement followed **breathing and relaxation exercises**. Oppenheim also recommends **gymnastic exercises**. **Hypnotism** and **suggestion** have been found to be of benefit in the cases complicating hysteria.

PROGNOSIS.—The prognosis is not favorable; the condition is chronic, and may last months and years. Strümpell has seen cases in which remissions occurred during pregnancy. Occasionally, recovery takes place spontaneously or after treatment, but when the disease has lasted for a long period, the ultimate outlook for permanent recovery is very poor.

Spasm of the Muscles of Mastication.—The spasm is limited to the muscles supplied by the motor fifth cranial nerve; it may be tonic or clonic. When *tonic*, the jaws are clenched and the mouth cannot be opened actively or passively, thus resulting in a general failure of nutrition. This variety of spasm is known as "trismus," and it occurs in tetanus and meningitis, less commonly in tetany and occasionally, temporarily, in epilepsy. Diseases of the pons and beginning acute bulbar palsy may be accompanied by trismus. It is rarely an isolated symptom. It is usually of reflex origin due to disease in the temporo-maxillary joint, or to unerupted teeth. It may also occur in hysteria.

When the twitchings are *clonic* they appear as rhythmical movements of the lower jaw, usually in the vertical direction; the movements may be so violent as to cause the teeth to chatter as in rigor (chills). These are seen during general epileptic and hysterical convulsions, and during the onset of febrile diseases, infectious in nature. As an isolated symptom, except transiently in hysteria, it is very rare. The grinding of the teeth in nervous children and adults during sleep is considered a variety of this form of spasm. This form of spasm is also observed in trigeminal neuralgia, and as a reflex in diseases of the ear. Schwartz and Burnett have observed by otoscope the spasm in the tympanic membrane.

TREATMENT.—The treatment consists in the **removal of the cause** and in relieving the symptoms. **Psychotherapy** is indicated in the hysterical cases.

PROGNOSIS.—The prognosis, if not due to organic lesions, is favorable; it usually disappears within a few weeks or months.

Spasm of the Tongue.—**SYNONYMS.**—Glossal spasm, Spasm of the hypoglossal region.—The tongue may participate in the spasms of chorea, epilepsy and hysteria, but isolated spasms of the tongue are rare. When they do occur independently, they are tonic, clonic, or mixed. They may interfere with chewing, swallowing and speech, the latter resulting in "aphthongia," a variety of stuttering.

ETIOLOGY.—The most common causes are emotional excitability, and reflex irritations from the throat, bad teeth and stomatitis. Strümpell mentions a case of spasm of the tongue in a glass blower. A neuropathic condition predisposes to the neurosis. Tongue "chewing" is frequently observed in infants during dentition, and in mentally defective children.

SYMPTOMATOLOGY.—The spasm is rarely continuous; it comes on in paroxysms every few days or weeks, occurring from 20 to 30 times a day. It is rarely seen during the night. An attack may last a few seconds or minutes, or may continue for hours; it does not always cease during sleep. B. Myers³ records an interesting case of tongue "chewing" which occurred in several members of the same family. The twitchings are sometimes preceded by paresthesiæ in the mouth and tongue.

TREATMENT.—The treatment consists in the employment of ordinary measures to maintain the patient's general health, the **removal of irritations**, the use of **sedatives**, the **application of galvanism** (the anode to the hypoglossal nerve), and **psychotherapy**. **Surgical measures** are hardly ever indicated. Lange stretched on one occasion the hypoglossal nerve, then resected it and finally had to divide the geniohyoglossus muscle.

PROGNOSIS.—The condition may last for years, but recovery ultimately takes place.

Spasm of the Muscles Supplied by the Glossopharyngeal Nerve.—

SYNONYMS.—Deglutition spasm, Pharyngismus.

OCCURRENCE.—This form seldom occurs spontaneously. It is usually

seen in hysteria and organic nervous disease, such as bulbar gliosis, in the form of tabetic crises, or in tetanus.

Spasm of the Muscles of the Neck.—SYNONYM.—Torticollis.

ETIOLOGY.—Torticollis affects mostly individuals of a neuropathic or psychopathic constitution; it is frequently associated with the neuroses and psychoses, and the patients very often present stigmata of degeneration. Rheumatic torticollis and congenital wry-neck are not included in this form of spasm, nor is the spasm of the muscles of the neck due to trauma or reflex causes, such as diseased cervical vertebræ, included under this heading. Peripheral irritation, even of the slightest degree, such as the wearing of a tight collar, may in predisposed individuals give rise to spasm; spastic torticollis may also be seen in diseases of the ear or brain.

Chronic poisoning with alcohol, lead or mercury is an important etiological factor. The condition may follow malaria, influenza, typhoid fever, pneumonia, chill, trauma, exertion or overstraining of the eye muscles from an error of refraction. In infants, rickets is a common cause. Brissaud considers this form of spasm purely psychogenic, and Oppenheim fully agrees with him when he says "the primary cause in typical cases is the neuropathic or psychopathic diathesis, and given this constitution, a number of factors, mental excitement, trauma, and overstrain of the cervical and nuchal muscles may bring on the spasm." The same author believes that the muscles of the neck are, next to those of the face, most commonly implicated in the movements of expression, and that localized muscular spasm frequently owes its origin to the fact that some emotional process, instead of becoming fixed in the mind as an imperative recollection, immediately invades the motor sphere and discharges itself in the form of a motor action—a spasm.

SYMPTOMATOLOGY.—Spasm of the muscles of the neck may be unilateral or bilateral; it may be limited to one or to several muscles of the neck. The muscles most commonly affected are those supplied by the spinal accessory nerve; the superficial or deep muscles or both may be affected; the sternocleidomastoid, however, is involved in most of the cases. The spasm may begin in one muscle and in the course of time spread to the others.

The spasm may be tonic or clonic, or both. Prolonged tonic spasm of the sternocleidomastoid is, as a rule, due to rheumatism, diseased vertebræ, or a congenital shortening of the muscles. The localization of the spasm in different parts of the same or other muscles will result in corresponding faulty attitudes. In severe cases, and at the height of the attack, the spasm may extend to the muscles of the trunk or extremities, simulating almost an attack of grand mal. When the spasm consists of "nodding" movements, it is called "spasmus nutans" or "salaaming spasm"; this form is most commonly seen in children during dentition, and may occur only during the night, or when the child is asleep. Involvement of the inferior oblique muscle results in simple "rotation spasm" (tic rotaire).

The spasms may be so slight as to be hardly noticeable, and of 110

inconvenience to the patient, or they may be so severe that speaking, eating and sleeping may be impossible and in these severe cases the involved muscles may, as a result of overactivity, become hypertrophied. Emotion, self-observation, and attempts to check the movements aggravate them; physical and mental rest, and the distraction of the attention alleviates them. On lying down the spasms cease at once, to return as soon as the patient gets up. Brissaud has pointed out that some patients are enabled to arrest the spasm by placing and holding a finger on the chin.

The spasms occur in paroxysms, but in some cases they may be continuous, at the rate of from 10 to 30 contractions per minute. Remissions and exacerbations are quite common. Except for the discomfort felt at the neck just before and during the spasms, there are no subjective or objective sensory disturbances, and there are no paralyses. In some cases, the spasms occur only during the night, but in the severe cases, even in the non-nocturnal ones, they may persist during sleep.

The psychic symptoms are those of the individual psychosis or neurosis, from which the patient may be suffering. Oppenheim saw a case in which the spasms of the muscles of the neck alternated with hallucinatory confusion. Gowers had a case in which the patient had melancholia for ten years before the onset of the spasm. In alcoholics, the spasms may be associated with delirium. Duchenne and others have recorded cases in which the condition was associated with writer's cramp.

DIAGNOSIS.—The condition is differentiated from *rheumatic torticollis* by the absence of pain and tenderness in the cervical muscles; from congenital torticollis, by the history of onset and the absence of changes in the cervical spine and in the muscles themselves. *Organic brain disease* will be excluded by the mode of onset, the presence of paralysis, and positive eye findings. At times it is almost impossible to differentiate this form of spasm from *general tic*, especially when the latter is not accompanied by "echolalia" (see Tics), or systematic, purposive movements; prolonged observation will in many cases make the diagnosis clear. It is well to bear in mind that general tic is usually preceded by a tic of the facial muscles, particularly the orbicularis palpebrarum. It is distinguished from *myoclonia* in that the latter is not limited to the muscles of the neck, and that the contractions are of lightning-like rapidity, and do not produce movements of the part. In *chorea*, the movements are irregular, the involvement is general, and the onset gradual. Its association with rheumatism and its manifestations is also diagnostic; but when the spasms coexist with chorea, the diagnosis may be impossible.

TREATMENT.—**General hygienic measures, light nutritious diet, a change of climate and occupation** with the removal of any discoverable causes are indicated. For the relief of the spasms, sedatives such as **bromids, gelsemium**, etc., are employed; the condition being a chronic one, the use of opiates and other habit-forming drugs is not advisable.

Orthopedic Measures.—The application of pads to press the head in

the direction opposite to the spasm, not too tightly applied, may be of use.

Electricity.—If pressure points can be elicited, the anode should be applied to them; if not, the anode is placed over the spinal accessory nerve, and the cathode over the muscle. **Faradism** has also been employed with good results, and **massage** properly directed may be of benefit.

Gymnastics.—Systematic exercises in fixing the head may be used as adjuvants.

Isolation, Psychotherapy, etc.—**Isolation** and **psychotherapy**, in the form of hypnotism, are of value in cases in which mental symptoms are a prominent factor. The "**inhibition treatment**" is highly recommended by Oppenheim and others. **Counterirritation**, in the form of **blisters** and the **cautery** to the **nape** of the **neck**, has resulted in marked improvement, and in some cases in permanent recovery.

Surgical Measures.—Section of the tendons, or section and stretching or resection of the spinal accessory nerve have been resorted to with varying results. Kocher and Quervain performed a total division of the tendons of the nape muscles, with more or less success in a number of cases. Brissaud is opposed to any surgical interference. At best, the improvement following surgical procedures is only temporary. No operation is to be performed unless the milder methods have been employed and have given no relief. Oppenheim ascribes the good results of surgical intervention to its effect as a counterirritant and its influence on the mind. Various forms of injections into the muscles have been employed, with results lauded by those who originated them.

PROGNOSIS.—The spasms may last for years or for a whole lifetime. They may attain a certain degree of severity and remain stationary. Usually they vary in intensity. Spontaneous recovery, or under treatment, may occur in the milder cases, especially when a removable cause can be discovered. In some few cases, the spasms may be very distressing and so severe that the afflicted individuals commit suicide.

PATHOLOGY.—No changes have been found in the central or peripheral nervous system. Some think the condition is due to involvement of the cortex, others to the cerebellum, and still others to the nerve nuclei of the affected muscles. The fact that so many cases yield to psychotherapy lends great support in favor of the psychogenetic nature of the condition.

Spasm of the Muscles of the Trunk and Extremities.—Spasm of the muscles of the trunk and extremities, not due to organic disease of the brain or cord, is rare. When such "**idiopathic**" spasm does exist, it may involve a single muscle, such as the rhomboid, the levator anguli scapulæ, the deltoid, the latissimus dorsi, the pectorals, etc., on either side or the same muscle on both sides, or a group of muscles innervated by the same nerve or by the same pair of nerve roots. When the spasm involves the lower extremities, the muscles of the calves are most frequently affected, although the muscles of the hip, the extensors or flexors

of the leg and foot, and even the cremasters and dartos have been found to be affected with spasm.

The spasms may be tonic or clonic, and may be due to endogenous or exogenous toxic conditions, infectious diseases, chill, trauma, mental or physical exhaustion, painful joints, amputation stumps and other reflex causes. The majority of those who suffer from these spasms are neurasthenic or psychasthenic individuals.

TREATMENT.—The spasm of the calf muscles is perhaps the one which is the most amenable to treatment. The treatment does not differ from that of the other varieties of spasm.

PROGNOSIS.—The prognosis as to permanent recovery is not very good; the spasms may last for months and years. They are exceedingly obstinate affections, and have a great tendency to recur.

Spasm of the Respiratory Muscles.—*Tonic spasm* of the diaphragm is occasionally met with in hysteria. The patients have a feeling of suffocation; the movements of the diaphragm cease during breathing, and this may be followed by acute pulmonary emphysema. Spasm of this muscle may also be met with in tetany and tetanus.

These cases are best treated by **cold douches**, while in a hot bath, with **hot fomentations applied to the pit of the stomach, electricity to the phrenic nerve** and **sedatives**. Obstinate cases may require **morphin hypodermatically**, and at times **general anesthesia with chloroform or ether** may be necessary to relieve the spasm.

Clonic spasm of the diaphragm is much more common than tonic spasm. The ordinary hiccough (singultus) is a good example of this form of spasm. It may, in severe cases, be so frequently and rapidly repeated as to interfere with speech, respiration and the taking of food. When it is a symptom of hysteria it is almost intractable. It may be due to reflex irritation from the gastro-intestinal or genito-urinary tract, or to undue emotion or excitement. Direct irritation of the phrenic nerve may also produce it; Strümpell mentions a case of mediastinal pericarditis characterized by this symptom. It is a grave symptom, commonly seen before death in organic brain disease, and in severe infections, especially of the peritoneum.

Spasmodic yawning (oscedo), spasmodic sneezing (ptarmus), spasmodic snoring (rhoncho-spasm), are seen from time to time in hysterical and other neurotic individuals. Spasmodic yawning may be an aura of an impending epileptic attack, and is sometimes seen in organic disease of the brain, particularly the cerebellum. Spasmodic attacks of coughing when not due to laryngeal crises or disease of the external ear, throat, nose and abdomen are usually hysterical in nature.

Saltatory Reflex Spasm.—**SYNONYM.**—Static reflex spasm.

This form of spasm was first described by Bamberger. As soon as the patient puts his feet to the ground, he begins to dance and to jump around on account of the clonic contractions of the muscles of the calf. The dancing, as well as the spasms, disappear as soon as the patient resumes the recumbent position. In some cases, the mere touching of the soles of the feet may bring on the spasms. There may be no

other symptoms than those of general nervousness or hysteria, although Erlenmeyer and Kast report that they have found in these cases the superficial as well as the deep reflexes exaggerated. The disease occurs in both sexes and at any age; it may appear spontaneously or may follow any of the infectious diseases. It has been known to occur as an occupation-neurosis in ballet dancers. Oppenheim considers it a symptom or a rare form of hysteria.

TREATMENT.—The *treatment* consists of **sedatives, electricity, wet packs and psychotherapy.**

PROGNOSIS.—The prognosis of this form of spasm is good; as a rule it does not last longer than a few weeks or months.

Camptocormia.—**HISTORY AND DEFINITION.**—Souques⁴ and Mme. Rosanoff-Saloff⁵ have described a form of “neuropathic pseudocontracture” of the muscles of the trunk, which they observed as one of the war neuroses in the last war. They called the condition “camptocormia” (formed from the words meaning “bending forward”).

ETIOLOGY AND SYMPTOMATOLOGY.—The condition is a functional one and is similar to normal bending forward of the body, except that the head is kept extended for the purpose of enlarging the field of vision. Except at the outset of the disease, walking is not interfered with, and the patients can readily bend down to pick up objects from the ground. They are unable to stand erect, and any attempt to straighten themselves out is followed by a prolonged tremor in the legs, but on lying down they can readily straighten and even hyperextend the back. The vertebral spines are neither tender nor painful, but the lumbar muscles may be tender to pressure. Neurological and x-ray examination is negative.

Almost all of the patients observed were victims of so-called “shell shock,” and although not actually wounded, they had been knocked down after an explosion with more or less loss of consciousness. The authors think that the condition is a neurosis to which neuropathic individuals are greatly predisposed.

TREATMENT.—For the treatment of the condition, Souques devised a **corset** which was applied when the patient's back became straight on lying down; those individuals who could not straighten their backs, even when lying down, had to be anesthetized. Aside from this, **measures ordinarily employed in the treatment of the neuroses** were resorted to.

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LOCALIZED AND GENERAL TIC

Synonyms.—Tic general, *Maladie des tics*, *Erinnerungskraempfe* (Friedreich), *Maladie des tics impulsifs* (Marina-Jolly), *Myospasia impulsiva*.

Etiology.—Most neurologists believe that tic is of cortical and not reflex origin. It appears, according to Oppenheim, most commonly between the ages of 7 and 15 years, affecting chiefly hereditarily neuropathic individuals; in isolated cases a direct hereditary transmission may be traced.

Symptomatology.—The condition begins with twitchings in the muscles of the face and neck; to these twitchings are gradually added movements that are apparently purposeful, such as throwing back the head, wriggling the shoulders, reaching or scratching the nose or beard, lifting up the collar, jumping or dancing, etc., all of these movements being repeated again and again in a most stereotyped manner. There may be a smacking of the lips, or a sniffing or a sucking movement. The movements pass off as suddenly as they come; distracting the attention usually arrests the movements, so that the patients can attend to their occupations or business. However violent a tic of the right arm or shoulder may be, the patient's handwriting shows no abnormality. As Patrick¹ has pointed out, when the impulse to tic can no longer be suppressed, the patient takes his pen from the paper, executes his tic and resumes the writing.

Attempts at too violent suppression of the tic aggravate it; the same may be said of physical and psychic excitement. Some patients suffer from obsessions and compulsory actions, such as counting the steps when walking, or counting the windows of the houses which they are passing. Others utter meaningless words or repeat the same words and sounds—*echolalia*; or use obscene words—*coprolalia*; or imitate movements—*echokinesis*. Individuals afflicted with a tic are usually absent-minded, and cannot concentrate their attention for any reasonable length of time. This is why children so affected are such poor scholars; they all lack full control of their will power. Pain is never an accompaniment of tic.

Differential Diagnosis.—Tic is characterized by frequent explosive repetitions of the same motor act, and is distinguished from simple reflex movements or movements of expression by the rapid, short, brusque and forcible nature of the movement, by their constant and frequent repetition as well as by their lack of purpose.

So many other diseases simulate the condition that the differential diagnosis may at times be very perplexing. It is distinguished from *chorea* by the fact that in the latter the movements are continuous, purposeless, general and irregular. A choreic never performs the same movement twice in the same way; a tiquer always performs the same movement in the same stereotyped manner. In *chorea* there are no periods during which the patients are free from the movements. The mental state of a choreic is entirely different from that of a tiquer; echolalia and coprolalia are never seen in *chorea*. In *hysteria* the twitchings appear suddenly after some excitement and there are usually other evidences of hysteria, such as paresthesiæ, anesthesiæ, etc., and the movements are not as stereotyped as in tic. The differentiation from the *myoclonias* may at times be almost impossible (see section on *Myoclonias*). In some of the *psychoses* there may be grimacing of the face and wriggling of the shoulders or limbs at the onset or at the height of the disease, but the history of the case and prolonged observation will clear up the diagnosis. The differentiation from *spasm* is discussed in the section on Spasms.

Varieties.—A proper scientific classification of tics is almost impossible, because the different varieties merge into each other; at times it is difficult to differentiate local from general tics, or to differentiate tics from reflex spasms and allied involuntary movements.

Tic may consist of a single movement—an isolated tic—or of a number of different movements which take place at the same time or in rapid succession, or it may appear in the form of a general tic. There is usually a "plurality" of movements involving several muscles supplied by different nerves. Thus, we have a "sucking tic," a "snuffing tic," a "licking tic," a "biting tic," "a grinning tic," "a scratching tic," a "nodding tic," a "gulping tic," etc. Under this heading of tics and habit spasms may be included the neurodermatoses, such as "tic de l'épilation of Raymond," called by Besnier "tic trichomaniac." Others call this form of tic "trichokryptomania," "trichotillomania" and "trichorrhexomania." These are all similar morbid states in which the affected individual has an ungovernable desire to pull one or more of his own hairs from the scalp, eyebrows, eyelashes, beard or mustache. There is a similar neurodermatosis, called "dermatothlasia," in which an apparently normal individual has a constant and uncontrollable desire to rub, scratch or irritate the skin in one or several parts of his body.

Curious tics are described by neurologists, who have had opportunities to observe some of the war neuroses. Mott reports the case of a pugilist who was terrified by the explosion of a bomb dropped by a Zeppelin, and who developed jerky purposive movements of the shoulder and head as if to avoid a blow, and facial grimaces such as a pugilist might assume in a fight.

Treatment.—Drugs seem to have no effect, though bromids may alleviate the spasms. When the patients cannot rest on account of the movements, chloral or chloroform inhalations may be employed. Wagner (cited by Oppenheim) reports thyroid extract of value in a few cases.

Light hydrotherapy (wet packs) and as much **rest and isolation** as possible are indicated. **Relaxation exercises** which keep the body and each part of it at rest, for short periods at the beginning and gradually lengthened as the treatment goes on, and **exercises** with a view to strengthening the power of inhibition, have been originated by Meige and Feindel and independently by Oppenheim with good results. The former make their patients drill before a mirror and call the exercise "psychomotor discipline." Pitres and Cruchet, Grossman and others recommend carefully directed breathing exercises. Clark and Oberndorf report cases of tic cured by **psychoanalysis**. On the whole, it may be said that true tic is more often benefited by **treatment properly directed**, such as **exercises of "control,"** and by **due attention paid to the psychic elements of the case** than by any other means.

Course and Prognosis.—The condition is a chronic and progressive one, but complete recovery may take place in cases of even long duration.

Historical Summary.—The term "tic" is adopted from the French. The condition was recognized by Friedreich and also by Charcot and his pupils, particularly Gilles de la Tourette² and Guinon.³ Further notable contributions on this subject came from Brissaud and his pupils, Meige and Feindel. The French school defined tics as "physiological acts, originally purposeful, but which have become acts apparently purposeless and meaningless." Oppenheim defines it as a "reflex, defensive or voluntary movement which has assumed an imperative character." The French investigators considered tics as psychoneurotic manifestations in individuals with infantile minds, in regard to their emotional reactions.

When the freudian school began the study of the psychoneuroses by psychoanalysis the mechanism of tic naturally attracted its attention, and it evolved the theory that a tic generally represents a purpose, that the purpose had been suppressed, and that the apparently senseless movement, when resumed, was a defense compromise which gave relief to the patient. L. Pierce Clark⁴ from a psychoanalytic study of three stubborn cases of "mental torticollis" is inclined to emphasize the auto-erotic gratification unconsciously afforded the tiquer by the tic, and to regard the aspect of the tic as a defense compromise of secondary importance.

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OCCUPATION NEUROSES

General considerations, p. 582—Etiology, p. 582—Symptomatology, p. 582—Diagnosis, p. 582—Treatment, p. 583—Course and prognosis, p. 583—Pathology, p. 583—Bibliography, p. 583.

Synonyms.—Occupation spasms, Occupation cramps, Coördinated occupation neuroses.

General Considerations.—Occupation neuroses are characterized by cramp-like contractions in the muscles, which appear only during certain definite complicated movements which have been acquired by habit or practice, whereas all other actions performed by the same muscles are normal.

In tailors and seamstresses, the spasms affect the muscles of the thumbs and forefinger. Telegraphers, pianists, cigar makers, shoemakers, blacksmiths, automobilists, milkers, barbers, drummers, tennis players, leather dressers, and others may be similarly affected. The lips may be involved in trumpet players; the vocal cords in singers; the eyes in watchmakers, microscopists and miners ("miner's nystagmus"); the calf muscles in dancers.

Writer's cramp (graphospasm) is the most common neurosis of this group. The onset is gradual; the patient first becomes tired, losing the usual control of his pen; he does not write as rapidly nor as smoothly. After a time, owing to the spasm of the muscles, he finds that he grips the penholder too tightly, and that the writing appears irregular and incomplete. The muscles involved are the interossei, the thenar and hypothenar, the lumbricales, the flexors, and extensors of the fingers and wrist, and the pronators and supinators. The more he thinks about the condition, the worse it becomes. Some patients have a simple weakness in writing; the muscles have a normal muscular force except during writing, when they are paralyzed (paralytic form). A combination of spasm and tremor is not uncommon. There is also a sensory form (neuralgic), in which the pain felt during writing makes this process difficult or impossible.

Etiology.—Occupation neuroses are most commonly seen in adult males of nervous make-up, so that more than one member of a family may therefore be affected. The exciting cause may be strain, bad posture, badly constructed and faultily held tools or instruments. Unusual attention to the work on hand aggravates the neurosis. It is a curious fact that, in violinists' cramp, the left hand may be involved as well as the right hand.

Symptomatology.—The subjects of this neurosis frequently complain of paresthesiæ, local weakness, tremors and pains in the affected limb, but objective examination fails to show any changes in motility or sensation. The reflexes are exaggerated and vasomotor disturbances are not uncommon, but these are probably due to the fact that almost all of these patients are neurasthenics.

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CHAPTER XXI

DYSKINESIAS

(Disorders of Motility)

BY MOSES KESCHNER, M.D., LL.B.

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Introduction.—In this chapter we intend to discuss a heterogeneous group of nervous affections characterized by the presence of some form of disturbed motility due to abnormal movements associated with changes in muscle tonus.

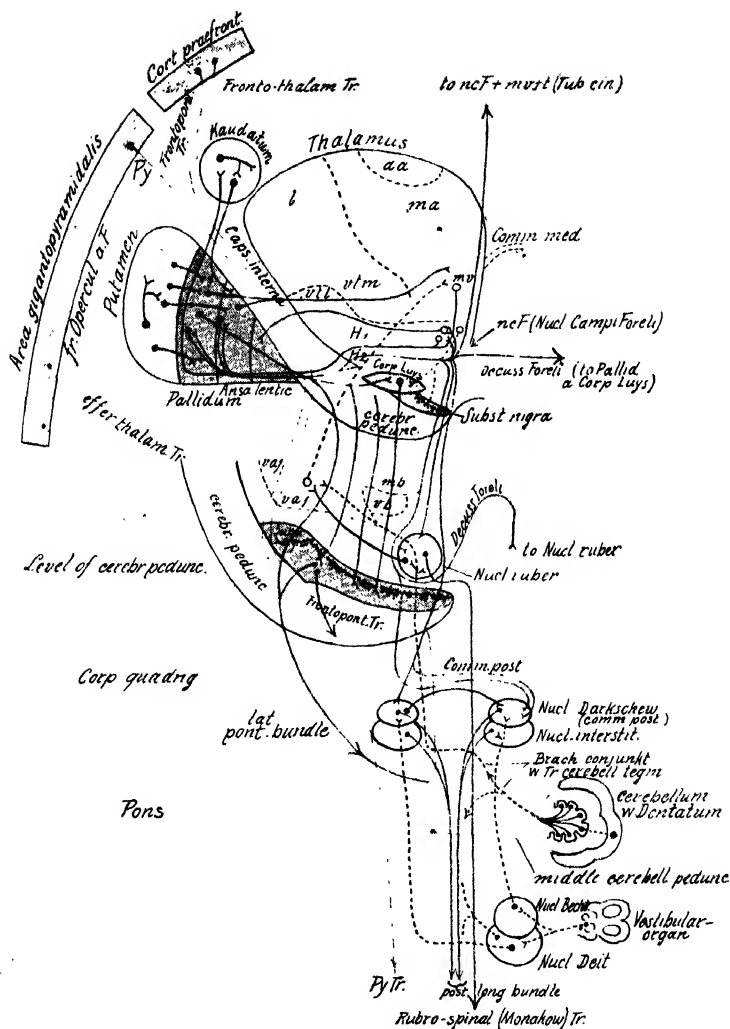
First we discuss the disorders of motility which in their purest forms are due solely to involvement of the extrapyramidal system (see below). In this category we include the tremors, the choreas, paralysis agitans, Wilson's disease and pseudo-sclerosis, the athetoses and the dystonias. Next we discuss the myoclonias with their characteristic lightning-like, rapid clonic twitchings of various groups of muscles; except for the myoclonias associated with epidemic encephalitis, which are also thought to be due to extrapyramidal involvement, there is perhaps no other justification for including the myoclonias in this chapter than the fact that abnormal movements constitute the pathognomonic phenomenologic (symptomatologic) feature of the condition. Next we discuss the various forms of spasm including occupational cramps and tics; most of the components of this group are either reflexogenous or psychogenous in origin; their inclusion in this chapter is justified principally on account of their close resemblance to the tremors and choreas. Next we discuss the different forms of myotonia; pathologically these are really types of muscular dystrophy (myopathies) but their principal clinical feature is a peculiar motor disorder—myotonia (see below)—whose pathophysiology cannot be satisfactorily explained on a purely myopathic basis; as a matter of fact, with the more recent methods of neurohistologic investigation, evidence is beginning to accumulate which is extremely suggestive that here too, neuronc factors—perhaps extrapyramidal influences—might play an important rôle. And, finally we discuss myotonia congenita (Oppenheim); this peculiar disorder of motility presents, pathologically, features of an atypical form of myopathy associated with a degeneration (embryonal defect?) of the anterior horn cells, and in some instances, of the peripheral nerves, i.e., a combination of myopathy, poliomyelitis and neuritis; clinically, it presents evidences

of disturbed (loss?) motility *without* paralysis or atrophy, and associated with a continuous hypotonia of the affected muscles. The inclusion of this condition in this chapter finds justification only in the fact, unscientific as that may be, that it is a disorder of motility associated with a change of muscle tonus, which on clinico-pathologic grounds cannot be classified otherwise than as a dyskinesia.

The writer recognizes the fact that from the point of view of classification, the physical arrangement of the material in this chapter cannot stand the critical test of any known scientific principle of classification of diseases, i.e., it is based neither on etiology, nor on pathology and pathogenesis, nor on hereditary or constitutional factors. It is merely a classification based on phenomenology (symptomatology). This is to a great extent due to the nature of the subject matter. None of the conditions discussed in this chapter can be regarded as definite disease entities; they are all merely symptom-complexes, whose etiology, pathology and pathogenesis have not as yet been definitely established. The writer is also aware of the fact that the title of the chapter "Dyskinesias" is also one that cannot escape just criticism. For with the broadest etymologic conception of the term "dyskinesia" in mind, this chapter should include every disease associated with a disturbance in motility; this would imply a discussion of every form of paralysis, central or peripheral, convulsions, tetany, tetanus, myasthenia gravis, Gerlier's disease, as well as disturbances of motility due to disease of the bones, muscles and joints. In conformity to the peculiarity of a cooperative method of text-book production and the difficulties of classification of the subject matter entailed by such method, as well as the necessity of avoiding repetition made it imperative to omit from this chapter many of the conditions whose symptomatology may be said to revolve around some form of disturbed motility, or dyskinesia, in the broadest sense of this term. For the discussion of these the reader is referred to the appropriate chapters in the text contributed by the other collaborators.

General Considerations.—Most of the motor disturbances discussed in this chapter are considered by modern neurologists as symptom-complexes or syndromes whose symptomatology is produced by pathologic changes involving the extrapyramidal system. The entire group of these syndromes has been designated by Strümpell as the "anystatic symptom-complex." The symptomatology of each component of this symptom-complex depends on the precise anatomic localization of the pathologic process in the extrapyramidal system, whereas the clinical course depends on the nature of the lesion, i.e., whether vascular, degenerative, inflammatory or neoplastic.

A great amount of work has been done in the last two or three decades on the anatomy, physiology, and pathology of the extrapyramidal system. Our knowledge of the pathophysiology of this system has been especially enriched by our experiences during the recent outbreaks of encephalitis. Although the problem is far from final solution, the writer is justified in giving a brief resumé of what is generally accepted as being the most rational view on the subject. The greater part of this resumé is based on the most recent conception of the anatomy and pathophysiology of the extrapyramidal system as elaborated by one of the most ardent students of this subject, Professor A. Jakob at the University of Hamburg.



Jason constructs three syndromes of extrapyramidal disease: (1) the choreiform, (2) the akinetic-hypertonic (or hypokinetic-hypertonic)—parkinsonism—and (3) the athetotic. These three syndromes have as their anatomic basis involvement of certain definite parts of the basal ganglions. The entire gray substance in this anatomic region is designated "the extrapyramidal system." In addition to the extrapyramidal system as represented by the basal ganglions there also exists a second mechanism subserving extrapyramidal motor coördination in the form of the fronto-ponto-cerebellar system; the former, however, is designated as the principal extrapyramidal system. This designation is only for the sake of brevity and is not meant to imply that the other extrapyramidal system is not as important as the principal one.

The principal extrapyramidal system consists of the striatum (caudate nucleus and putamen), globus pallidus, corpus Luysii and substantia nigra Soemmeringii. The red nucleus, an outpost of the cerebellum, indirectly also belongs to this system. The interrelations of these centers are best represented in the following schematic representation (Fig. 1).

An analysis of this diagram will show that aside from the substantia nigra, which also receives its impulses directly from the cortex, the extrapyramidal system receives impulses from the mid-brain, brain stem, and thalamus. The mid-brain is of great significance for the various metabolic processes of the organism and especially for the tonus of the entire musculature of the body. The thalamus seems to be the great collecting organ for proprioceptive and extrareceptive stimuli, as well as for cerebellar impulses, and stands in intimate afferent and efferent connection with the entire cortex. The thalamus is of great significance to the individual not only by informing him of changes going on within the body itself but also of its relation to the outside world. It is therefore an important organ as far as sensation, emotion, psychic tone and feeling in general of the individual is concerned. By apprising the individual of the relationship of his body to the outside world, the thalamus may also be regarded as one of the most important components of the coördinating mechanism of the extrapyramidal system. In this connection it must also be remembered that the chief function of the cerebellum—a proprioceptively stimulated organ—is to intensify and to check this entire coördinating apparatus especially in the matter of controlling specifically the synergic components necessary in the determination of the direction of movements of the entire body and parts thereof. It acts as a stabilizer for the motor apparatus of the mid-brain and brain stem. This it accomplishes to a great extent through the red nucleus.

Thus the extrapyramidal system is the efferent organ of the thalamus and hypothalamus, in which afferent stimuli are translated into highly developed motor and tonic phenomena. It is, therefore, a center for the movements of expression, flight, fright and defence; it is concerned with the regulation of movements for the automatic change of position and attitude and for associated movements. It is also of great significance for the "running out" of the various components of movement essential for orderly sitting, standing, walking, chewing, swallowing, talking, as well as for bladder control. It must also be regarded as part of the voluntary tract through which cortical impulses are directed to

the anterior horns. Bearing these facts in mind, it is obvious that a lesion anywhere in this most complicated system will give rise to disturbances in motility and to changes in muscle tonus. Diseases of the extrapyramidal system will, therefore, be manifested clinically by changes in muscle tonus and some form of disturbed motility without paralysis.

By far the greater majority of cases of extrapyramidal disease give rise to a more or less permanent increase in the tonicity of the skeletal musculature. An example par excellence of this form of hypertonicity is the so-called parkinsonian rigidity. This is characterized by an increase in the permanent, plastic, form-giving muscle tonus accompanying passive resistance to muscle stretching, delayed muscle after-contraction, adaptation and fixation tension giving rise to peculiar postures and attitudes. The rigidity is "cadaveric" and "waxy" and is not influenced by the speed of active or passive movements in an attempt to overcome it. During movement the examiner can usually elicit the so-called "cog-wheel phenomenon" which is not unlike the sensation experienced on "pulling a ratchet." This form of rigidity can readily be distinguished from the elastic springlike form of spasticity observed in pyramidal lesions. Another peculiarity of extrapyramidal hypertonicity is that in spite of its severity it may be overcome on repeated passive or active movement, except in advanced cases in which marked contractures have already resulted. Extrapyramidal rigidity affects flexors and extensors more or less equally. This is another important diagnostic point in differentiating pyramidal from extrapyramidal disease, because in the former there is, as Wilson states, "a selective accession of tone, as in the flexed arm of hemiplegia and the extended or flexed leg in paraplegia. No amount of bilateral cortico-spinal motor disease can produce hypertonicity in all muscles indifferently."

In some cases, and these are in the minority, the tone may vary alternately from hypertonicity to hypotonicity, giving rise to a condition designated as "dystonia." Here hypertonus follows temporary hypotonus in a confusing and irregular fashion. A segment of a limb, or an entire limb, or several limbs and in advanced cases even the entire body contracts involuntarily, in one moment, in flexion, and in the next, in extension. This variation in tonus gives rise to most bizarre twisting and torsion movements and attitudes.

The involuntary movements of extrapyramidal disease may roughly be divided into two groups: (1) The akinetic and hypokinetic and (2) the hyperkinetic.

The hypokinetic and akinetic phenomena are attributed to a diminution or failure of innervation due to a lack of stimulation from the extrapyramidal system and of discharges from its centers. The components of rigidity associated with these phenomena are due to a disturbance or loss of the regulating function of the hypothalamic centers, the motor apparatus of the brain stem and of the cerebellar stabilizing function through the extrapyramidal system. These phenomena are clinically characterized by a slowness and poverty of movement especially noted during the initiation of movement, and failure to obtain the necessary amount of smoothness or gracefulness of movement during its execution. This is best exemplified by the mask-like facies, the loss of facial expression and the diminution or loss of the physiological associated movements during walking, making a fist, etc.

The hyperkinetic phenomena are the well-known choreic and athetoid movements, torsion spasm, hemiballismus (or throwing movements), myoclonias, ties and tremors. In the study of the hyperkinetic phenomena it must be borne in mind that a normal cortico-spinal system cannot inhibit the effects of striatal disease; it cannot inhibit the hyper-tonicity and hyperkinesia of pure striatal disease, because the striatum is in a sense an autonomic organ into which centrifugal fibers from the cortex do not enter.

Extrapyramidal disease gives rise to various forms of "shaking movements" or tremors. The commonest of these is the "pill-rolling movement" of the fingers observed in paralysis agitans. If this movement be suppressed in one extremity it will appear in another, or if present in one or more than one, its suppression in any one of them will intensify the movements in the others ("overflow of the tremor"); emotion increases the tremor. As the disease advances and the limbs become fixed by rigidity the severity of the tremor is greatly lessened. The tremor persists but is diminished during rest ("tremor of repose"). Hunt regards tremor as a release phenomenon resulting from the loss of striatal control, and the expression of spontaneous activity in certain infra-striatal centers of the extrapyramidal system. It is, therefore, a disorder of motility and, in a sense, analogous to chorea and athetoid movements. Hughlings Jackson, however, believes that tremor is the result of rigidity and defines tremor as "rigidity spread thin." The paralysis agitans (parkinsonian) tremor is distinguished from pure cerebellar tremor by the fact that the latter is a so-called "intention tremor" which is associated with dyssynergia, dysmetria, dysdiadokokinesis and ataxia. Cerebellar tremor is never a tremor of repose; it is coarse, arrhythmic, beginning with movement and increasing as the object is reached. The parkinsonian tremor is a fine rhythmic tremor diminishing in severity and usually ceasing when the goal is reached. Occasionally one observes in extrapyramidal disease a combination of both of these tremors. This form of tremor Hunt has designated the "striocerebellar tremor." It is usually found in lesions in the region of the mid-brain which is the meeting place of the efferent systems of the corpus striatum (pallidal system) and cerebellum (dentate system) which converge and terminate in the red nucleus system. In other words, it is a tremor of the strio-rubro-spinal system.

Strümpell attributes tremor in general to a failure of reciprocal innervation during which the muscle groups fixing a joint are not innervated simultaneously, giving rise to an oscillatory innervation of the joint. This explanation, however, would not explain the tremor at rest. Marburg believes that tremor is due to a simultaneous disturbance of tonus and faulty innervation of the antagonists resulting in a pendulous movement of the joint, and should the innervation of the antagonists be so delayed that a slight contraction is followed by a marked relaxation, the movement becomes a "throwing," choreiform, and even an athetotic movement. Marburg, therefore, regards choreiform movements as tremors of unusually wide excursion and unequal tempo. Wilson, on the other hand, associates tremor with the play of both striatal and cerebellar impulses on the mesencephalon, and he holds that in the former the tremor is more apt to be accompanied by rigidity, whereas in the latter it is apparently associated with hypotonicity.

On the basis of these anatomic-physiologic considerations and disturbances of function in clinically observed cases subjected to exhaustive histopathologic investigation, Jakob gives the following views concerning the functions of the individual components of the extrapyramidal system: The *striatum* is the true center for gestures of expression and for reactive and defense movements; it also gives tone to the *pallidum*. The *pallidum*, which in the newborn is the center for primitive and incoördinated, automatic movements, becomes in the adult the center for locomotor synergies of single muscle groups and of segments of the extremities in the service of striatal locomotor automatic acts, which are built on the locomotor synergies of the cerebellum and brain stem. In addition, it also plays a significant rôle in the tonus-regulating mechanism. The *corpus Luysii* apparently contrasts the locomotor synergies of entire parts of the body, with special emphasis on cerebellar balancing components. The *substantia nigra* is a tone-controlling center which, in a special way, serves to determine the orderly succession of movements. The *striatum* and *pallidum* are somatotopically united, and like the *corpus Luysii* innervate both halves of the body, but more the contralateral half. Similar relations apply also to the *substantia nigra*. Finally, the whole extrapyramidal system is functioning when the *thalamus* is active, and as a result of these activities the pyramidal system has the most favorable condition for its own functioning, so that the anterior horn cells are constantly receiving normally directing impulses through the influence of the extrapyramidal system.

TREMORS

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General Considerations.—The term tremor is applied to regular, involuntary, alternating movements taking place in smaller or larger excursions, always in the same plane, produced by involuntary contraction of certain muscles and their opponents.

A *simple tremor* is one affecting a single muscle group and its antagonistic group. A *compound tremor* is one in which several groups of

muscles and their opponents are in action and produce a complex movement, i.e., flexion and extension of the fingers combined with pronation and supination of the forearm. A *static tremor* is one which appears only during active movement, when the involved limb is held at rest in a special position. A *motor tremor* appears only during the act of movement of the involved limb, but not when the latter is held at rest.

Depending upon the nature of the disease producing the tremor, it may appear only during rest, as in parkinsonism (*paralysis agitans*), or during action or "intention," as in multiple sclerosis. In some cases tremor will appear only when the patient is under the influence of emotional excitement, such as pleasure, fear, or anticipation. Tremor may appear in paroxysms as in the neuroses, or it may be present continuously as in organic disease of the nervous system or in chronic toxic states.

In studying tremors, it is important to determine whether they are intensified or diminished during action or during rest, and what effect supporting the trembling limb has on the intensity of the tremor; also what effect self-consciousness and emotional states have on its intensity and amplitude.

Special attention must also be paid to the rapidity and regularity or rhythm of a tremor, so that we may be able to distinguish between rapid and slow tremors. Tremors are considered rapid, when they consist of from eight to ten oscillations per second, and slow, when from three to five oscillations per second; occasionally tremors are encountered, which, as far as rapidity is concerned, stand midway between the two. Tremors may be fine or coarse; a rapid tremor is usually fine in its oscillations, and is known as a *vibratory tremor*.

When there is difficulty in eliciting a tremor during simple acts of movements, it may be brought out when the patient's finger or toe is allowed to follow the examiner's finger while the latter is being moved away from the limb which is being tested.

Nystagmus is a variety of action or intention tremor of the extraocular muscles. (For the mechanism and significance of this abnormal movement, the reader is referred to the chapter on the Cranial Nerves and their Diseases, cf. p. 19.) A contraction and undulation which rapidly passes from one muscle bundle to another in the same muscle, so that the entire muscle appears as if a wave passed over it, is a *fibrillary tremor*. This form of tremor or twitching is seen in incompletely atrophied muscles following disease of the anterior horns, such as amyotrophic lateral sclerosis, or in peripheral nerve disease such as facial paralysis (Bell's palsy). Fibrillary tremors can be elicited by brisk percussion of the involved muscle or muscles; they never produce movement of a limb; they are intensified by exposure to cold and by excitement. Fibrillary tremors must be distinguished from the fibrillations seen in delicate and nervous persons during exposure to cold and during physical overstrain without the presence of organic nervous disease.

Transient quivering of a muscle affecting a few muscle bundles of one muscle without producing movement is called *myokymia*—popularly known as "live flesh," and is seen in anemic, weak and neurasthenic individuals. Myokymia is not associated with atrophies and is not affected by rest or exertion.

Rhythmic variations in the amplitude of tremors have been observed

with a certain regularity in each "tremor movement," and such tremors have been designated as *allorhythmic tremors*. They have been observed in Basedow's disease, in metallic poisoning, paralysis agitans, alcoholism and in the neuroses. Most neurologists attribute no significance to this type of tremor.

It is well to bear in mind that healthy people have tremors under certain conditions such as excitement, chills, and violent physical exertion. Pitres calls these tremors *physiological tremors*. Smoking and drinking to excess may produce a transient or persistent tremor. Tremors are occasionally observed during the onset of infectious diseases and during convalescence from severe and exhausting diseases. Tremors must not be confused with the fine clonic movements noted in very spastic limbs on attempting to elicit the deep or tendon reflexes.

Varieties.—TREMOR IN ORGANIC NERVOUS DISEASE.—(1) *Post-hemiplegic Tremor*.—This variety of tremor is observed in Benedikt's syndrome (see chapter on Hemiplegia, Vol. IX, p. 494, in lesions of the cerebral peduncle) of the frontal lobes, pons, corpora quadrigemina, red nucleus, basal ganglions and cerebellum. The tremor may assume the characteristics of a coarse "intention" or "action" tremor, as in multiple sclerosis involving the brain stem and the cerebellum, or it may be a fine "to and fro" tremor as in parkinsonism (paralysis agitans). Tremor is also a prominent symptom in Wilson's disease (bilateral lenticular degeneration), in Westphal-Strümpell's pseudosclerosis, and in dystonia. The character of the tremor will depend on the anatomic site of the lesion, and will be the same regardless of whether the latter be inflammatory (encephalitic), degenerative, vascular or neoplastic in nature. As a rule, tremor does not occur in a segment of the body that is completely paralyzed.

The tremor observed in *cerebral arteriosclerosis* may also be in the nature of an "intention" or "action" tremor, or it may assume the character of a parkinsonian (paralysis agitans) tremor, depending upon the precise anatomic distribution of the arteriosclerotic foci; in cases of advanced cerebral arteriosclerosis with diffuse lesions the tremor may be both parkinsonian and intentional in type. So-called *senile tremor* is probably a tremor due to cerebral arteriosclerosis. Tremor may also be a prominent symptom in *Friedreich's ataxia* and *Mariette's ataxia* and in other system diseases associated with lesions of the cerebellar mechanism. *Essential tremor* (névrose trémulante) is an expression of a degenerative process in the central nervous system, without other clinical evidences of a progressive nervous disease; hereditary factors apparently play an important etiologic rôle; most of these patients give a history of epilepsy, syphilis, alcohol and nervous disease in the antecedents. This variety of tremor may be local or general; it may appear at rest or during action (intention). It may begin at any age, and may be familial. Mitchell has described a form of hereditary tremor affecting mainly the head. The most common involvement, in the order of frequency, are the hands, the legs and the muscles of the face and tongue. It is occasionally associated with choreiform movements, tic and nystagmus. The tremor may last throughout life, with periods of remission. Total disappearance almost never takes place, and treatment seems to have little or no influence.

A localized or generalized tremor is occasionally observed in *epilepsy* immediately preceding or following an attack.

Tremor is a common and early symptom in *general paresis*. It may be confined to some particular muscles or to one side of the body, or it may involve the entire body. It is most commonly seen in the facial muscles, lips and tongue; it is readily brought out by asking the patient to speak or to show his teeth, or to protrude the tongue. The latter movement is carried out in a very characteristic manner; the tongue is thrust out, showing a very fine and rapid tremor coming in waves along its muscles, the mouth remaining wide open, or the tongue is repeatedly thrust out and drawn back. Tremor of the fingers is also very common in paresis, but is not as characteristic as that of the lips and tongue; it is fine and irregular, and may be slow or rapid; it is intensified by physical or mental excitement, it accompanies movement, and may persist during rest. The tremor in paresis may occasionally be associated with choreiform and myoclonic movements.

TREMOR IN INFECTIOUS DISEASES.—Tremor may be met with in the course of infectious diseases, especially the meningitides. Cases of *polio-myelitis* have been described in which tremor of the affected limb preceded the paralysis. Clement (cited by Oppenheim) saw a tremor, like that of paralysis agitans, in the course of *typhoid fever*. De Brun notes a tremor in every case of secondary *malaria*. In two of his cases, the tremor was exaggerated by an impending malarial paroxysm. He believes that in some cases the tremor is toxic in nature, and in others that some organic lesion of the brain is responsible for it.

Various tremors and twitchings have been observed during the recent epidemics of *lethargic encephalitis*. In some cases they occur in the prodromal period and persist throughout the entire disease; in others they appear during its course. In either case they may remain in evidence months and years after all other symptoms have disappeared. They may be localized in any part of the body, or they may involve the entire body. The tremor may be fine or coarse, regular or irregular, rapid or slow; it may persist during sleep. Most of the tremors are *parkinsonian* and a good many *myoclonic* in type. In some of the cases generalized or localized *choreiform*, *tic-like*, and *dystonic movements* were observed either alone or in combination with tremor, and in many cases the severity of the former overshadowed the latter.

TOXIC TREMORS.—Toxic conditions give rise to various forms of tremor. The most commonly observed toxic tremors are those due to the excessive use of tobacco, coffee and tea. These tremors are usually fine and very rapid in character; they are exaggerated by exercise and mitigated by taking food. Those addicted to opium, heroin and morphin show a general tremor of the entire body, especially when deprived of their usual daily allowance of their habit-forming drug. Oppenheim once saw tremor and lateropulsion in a gouty patient after colchicum poisoning and made the diagnosis of paralysis agitans, but the symptoms promptly disappeared after the amount of colchicum was reduced.

The most commonly observed toxic tremor is that occurring in *alcoholism*. It is a coarse tremor (6-9 oscillations per second) involving the fingers and tongue, although in severe cases it may also involve the limbs and head. It is most marked in the morning before the patient has had his breakfast, and is diminished or may even entirely disappear after the administration of alcohol. Quinquand has described a sign which he considers pathognomonic of chronic alcoholism. When the ex-

aminer presses the palm of his own hand, which is held in a vertical position at right angles to the patient's fingers, which are fully extended at the interphalangeal joints, and widely spread apart, nothing in particular will be noticed for the first two or three seconds, but after this, the examiner will feel a slight quivering, jerking or crepitation, as if the phalanges of each finger were knocking against the other, trying to reach the examiner's palm. The same phenomenon may be elicited in tremors due to other conditions, and it is doubtful whether it can be considered pathognomonic of alcoholism. Alcoholic tremor is very marked during an attack of delirium tremens, and persists long after the delirium has disappeared.

TREMOR IN BASEDOW'S DISEASE (HYPERTHYROIDISM).—Although tremor does not always persist throughout the course of *hyperthyroidism*, it is, nevertheless, such a constant symptom as to be considered pathognomonic. It is a fine, regular, rapid vibratory tremor most marked in the fingers and hands on extension, but may be seen and felt in the head, shoulders, trunk and feet. Irritation of any kind, cold, physical or mental excitement, aggravates the tremor, and although it is diminished during rest, it may in some cases be perceptible even then. Tremulous respiration, in which the tremor is synchronous with that of the outstretched hands, is, according to Purves Stewart, one of the most constant signs of Basedow's disease.

The close relationship existing between the functions of the thyroid gland and the adrenals has been utilized by Goetsch in the elaboration of a skin reaction which is diagnostic of hyperthyroidism. Accompanying the local reaction in the skin following the hypodermatic injection of a 1:1000 solution of epinephrin, there is an exaggeration of the tremor and the nervous symptoms in general. (For details of this test cf. Vol. VIII, p. 240.)

A generalized fine tremor is often seen in women during the menopause which is usually ascribed to hysteria or neurasthenia, but which will be found on careful observation to be due to thyroid dysfunction.

TREMORS DUE TO CHRONIC METALLIC POISONING.—Chronic poisoning with metals such as bismuth, copper, arsenic, manganese, lead and mercury also give rise to tremor. The tremor of mercury poisoning precedes the buccal symptoms and palsy, and involves the muscles of the face and extremities; it is usually constant, from 5 to 6 oscillations per second, coarse in nature, best elicited during movement, and increased during attempts at suppression; it is widespread and associated with muscular weakness, prostration, tremulous voice and mental deterioration. Treatment instituted for mercurial elimination is usually followed by improvement in the tremor and the patient's general condition. Chronic poisoning with carbon monoxid and carbon disulphid may also give rise to tremor. These tremors are due to actual lesions in the basal ganglions, especially the striatum.

TREMOR IN THE NEUROSES.—Tremor is a common symptom in *neurasthenia*. It is usually a rapid, fine, vibratory tremor accompanying active movements and intensified by physical overstrain and emotional excitement. It simulates the tremor seen in healthy individuals after excessive smoking, drinking and venery. In addition to this form of general tremor Oppenheim describes in neurasthenia a *fibrillary tremor* affecting the lids, lips, interossei and quadriceps femoris on exposing the leg; it may resemble *myokymia*.

The tremor characteristic of *hysteria* is one of large amplitude and of from 5 to 7 oscillations per second. It is intensified by mental excitement and voluntary movement, simulating the tremor of disseminated sclerosis in this respect, but differing from it in that it lasts longer than the movement which evokes it, or it may fail to appear in certain movements. It may be as marked during rest as during action; it is inconstant and *polymorphous* in type. It may appear in paroxysms, or it may be continuous; it may be very slight, or so marked as to develop into a clonic spasm. It may involve the legs so as to make locomotion impossible—or it may be most severe when the patient is on his back; it may affect any or all extremities or the entire body. Suggestion and *hypnotism* may diminish it or stop it. During the last war various kinds of tremors were encountered in soldiers suffering from so-called *shell-shock*. Some of the tremors simulated the tremor of multiple sclerosis, others that of paralysis agitans, and still others were polymorphous and difficult to classify. Patients suffering from *traumatic neuroses* and *malingerers* very frequently show "functional" tremors that present difficulties in differential diagnosis.

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THE CHOREAS

Synonyms.—Infectious chorea, Sydenham's chorea, Sydenham's disease, St. Vitus' dance, Chorea of childhood, Chorea minor, Danse de St. Guy, Danse de St. With, Myotyrbie (Dartigues), Periodical jactitation (R. Watt), Chorée, Veitstanz, Veitsdands, Vit-Tancz, Plasawicy scelotirbe, Corea, Folie musculaire (Bouillaud), Insanity of the muscles (Maudsley), Chorea Sancti Viti.

Introduction

The word "chorea" derived from the Greek (*χορεία*—dance) is suggestive of many different conditions which are all characterized by abnormal muscular movements, twitchings or spasms. The conditions in which such movements are the predominating symptom form a heterogeneous group, which includes infectious or Sydenham's chorea, the degenerative choreas (the Huntington group), the senile choreas, the symptomatic choreas, etc. The most common affection belonging to this large group is infectious or Sydenham's chorea.

SYDENHAM'S CHOREA

- Frequency, p. 474—Etiology, p. 474—Predisposing causes, p. 474—Associated diseases, p. 476—Rheumatism, p. 476—Endocarditis, p. 476—Bacterial invasion, p. 477—Tonsillitis, abscessed teeth, aural discharges and other focal infections, p. 478—Scarlet fever, p. 480—Whooping-cough, influenza, tuberculosis, typhoid

fever, gonorrhea, small-pox, chicken pox, diphtheria, cerebrospinal meningitis, pyemia, malarial poisoning, p. 480—Symptomatology, p. 481—Clinical history, p. 481—Physical findings, p. 481—Psychic states, p. 484—Laboratory findings, p. 484—Duration, p. 485—Recurrences, p. 485—Diagnosis, p. 485—Differential diagnosis, p. 486—Complications, p. 487—Clinical types, p. 491—Treatment, p. 492—Prognosis, p. 496—Pathology and pathogenesis, p. 497—Summary of etiology, pathology and pathogenesis, p. 499—Historical Summary, p. 499—References, p. 500.

Definition.—Sydenham's or infectious chorea is a disease occurring chiefly in children, due to some toxic or infectious agent, which acts on the central nervous system by producing irregular involuntary contractions of the muscles, resulting in purposeless movements, and associated with muscle weakness and mental irritability.

Frequency.—Sée, in a hospital experience of 22 years, saw 531 cases of chorea, approximately 0.8 per cent. of all his cases. Rufz found, among 32,976 sick children, 189 cases, about 0.5 per cent. Abt and Levinson²³ found, among 10,150 sick children, 226 cases of chorea. Wicke observed the disease only in 0.18 per cent. of his cases. So many cases of mild chorea are discovered accidentally, that statistics as to frequency have little or no value. There is no doubt, however, that chorea is a common disease of childhood.

Etiology.—PREDISPOSING CAUSES.—*Seasonal Influences.*—Weir, Mitchell, Siukler, Sachs and Starr found most of their cases to have begun during the spring. This is also the writer's experience. According to McCarthy of Philadelphia, the fact that most cases of chorea occur during the spring is most probably due to the poor general tone of the children after they have been confined indoors, and have been much occupied with their school work during the winter months. Morris J. Lewis¹ undertook a most elaborate inquiry, for a period of ten years, with a view of ascertaining whether there is any relation between temperature, humidity or barometric variations and the occurrence of chorea, but he could find none.

Sex.—The proportion of males to females is about 1:3. Starr found in 466 cases, 136 males and 330 females. The British Chorea Committee found in 436 cases, 114 males and 322 females. Sinkler collected 328 cases, of which 232 were females and 96 males. Gowers, who combined the statistics of several other authors with his own, found that of 1,000 cases, only 365 were boys. In Osler's cases the proportion was 1 male to 2 females. Abt and Levinson's²³ cases were in the same proportion. In the last 100 successive cases seen by the author there were 34 males and 66 females.

Age.—Friedländer saw congenital chorea in two sisters. Mayo and Sinkler each refers to a case of congenital chorea, due to a fright of the mother during pregnancy. Richter, Fox, Heller and others also report congenital cases. Haven has seen 2 congenital cases among 195 cases of the disease. Simon has met the disease in children a few days old. Sachs claims to have seen several cases in children less than one year old; the youngest case of Wicke was 2 years old, and of Holt 4 years. Statistical studies would seem to show that the greatest number of cases occur between the ages of 7 and 13, the period of most active growth, when the greatest demands are made on the nervous system, and when metabolism is most active.

Racial and Climatic Influences.—Climate seems to have little influence on the disease. According to Axenfeld, on the authority of Rufz de Lavison, the disease is unknown in Martinique, Guadeloupe and other hot climates. It is comparatively infrequent in our Southern states. Sachs claims to have seen in the New York Polyclinic a number of negro children with chorea, curiously all boys, in whom competition in school seems to have been the exciting cause. In New York City, children of German and American ancestry seem to be more frequently affected than children of Irish descent. It seems to be quite prevalent in Italy. Jews, with their natural tendency to nervous diseases, form a prolific source of the disease.

Hereditary Influences.—In the consideration of hereditary influences the relation of chorea to rheumatism on the one hand, and to other nervous diseases on the other, must be borne in mind. An analysis of the cases seems to show that direct heredity in chorea is very rare, but that individuals whose ancestors have been afflicted with alcoholism, lues, saturnism, epilepsy, hysteria, tics, insanity or other nervous diseases (i.e., those with a tendency to neuropathy or psychopathy), are more predisposed to the disease than others.

Social Influences.—Poor children, especially those living in overcrowded, ill-ventilated tenements, and those who are poorly nourished, seem to be most susceptible. Bright and ambitious school children seem to be more frequently affected than those mentally retarded.

Imitation.—The disease is said to be brought on frequently by "imitation." It is doubtful whether the epidemics of chorea observed in boarding-schools can be considered genuine chorea; the muscular twitchings seen in these patients are most probably hysterical in nature.

Toxic Agents.—Choreiform movements are frequent in the cerebral forms of chronic poisoning with lead, mercury and other metals. The excessive use of tobacco has been observed in young adults to be associated with irregular, involuntary, generalized, or localized twitchings. Symptoms similar to ordinary chorea have been noticed during intoxication with hyoscin, and other derivatives of belladonna; in these cases the choreiform movements are constantly associated with mild delusions. This symptom-complex is described in books as "*hyoscin chorea*." Poisoning with other drugs belonging to the vegetable group also gives rise occasionally to choreiform movements; but the symptoms are transitory and disappear promptly on the withdrawal of the drug. They can hardly be considered manifestations of chorea as described in this chapter.

Emotional Disturbances.—Fright, emotional excitement and general mental distress seem to play an important rôle as predisposing causes in the disease. All writers agree that if fright is to be considered a predisposing cause, the interval between the fright and the onset of the chorea must not exceed one week; the average interval is from 3 to 5 days. Most frequently, the chorea immediately follows the fright. In children who have had chorea, trivial occurrences, such as a thunder-storm or a severe scolding, may be sufficient to bring on a recurrence of the disease. *Overstudy* has undoubtedly much to do with the development of chorea, but the importance of *masturbation* as a cause has been overestimated.

Trauma.—Chorea may occasionally be traumatic in origin, but it is

then dependent upon some grave cerebral affection or meningeal irritation.

Reflex Irritation.—Reflex irritation is believed to play an important rôle in the causation of the disease. Errors of diet—especially an excessive meat diet—are often said to be responsible for an attack of chorea; in these cases there is frequently a previous history of broken sleep, night terrors, enuresis and somnambulism. Tardieu reports a case of chorea which was promptly cured after the expulsion of eight *Ascarides lumbricoides*; similar cases are reported by Hanfield-Jones and by Hamilton.

Ocular Defects.—Even low degrees of hypermetropic astigmatism have been ascribed as causal agents of chorea. Stevens claimed that with the correction of errors of refraction the choreiform movements had disappeared, but the New York Neurological Society after an impartial investigation of Stevens's claims came to the conclusion that the facts did not warrant their adoption.

Local spasms of the facial muscles, "the *habit chorea* of Mitchell," have been pointed out by Jacobi to be associated with enlarged tonsils and adenoid vegetations in the nasopharynx. These so-called habit choreas, however, are not, properly speaking, "choreas"; they are forms of facial spasms or tics.

Reflex irritations from the *genito-urinary tract* have, in the absence of any other ascertainable cause, also been considered etiologic factors in chorea. The fact that many cases of chorea occur at or near puberty, that it is more common in females than in males, and the peculiar course of the disease in the chorea of pregnancy are, in the opinion of some authors, arguments in favor of *endocrine dysfunction* as being an etiologic factor in the disease.

ASSOCIATED DISEASES.—In 1831, Thomson² described "metastatic rheumatic inflammation of the cord and its nerves" as the chief cause of chorea. Bright,³ in 1838, considered post-rheumatic heart disease as a link between chorea and rheumatism. Hughes, in 1846, found, out of 104 cases of chorea, that 89 had either rheumatism or heart disease. Roger, in 1866, attempted to establish the fact that rheumatism, heart disease and chorea are parts of the same clinical entity, and he differentiated between "rheumatism-chorea," "cardiac chorea" and "cardiac rheumatic chorea," depending upon which one of these affections manifested itself. Jacobi, in 1875, in his essay on "Rheumatism in Children" pointed out the close relationship existing between rheumatism, chorea and endocarditis and that usually *rheumatism* came first, later *endocarditis*, and last *chorea*. A reverse of this order he considered to be an exception and even then he had his doubts, because rheumatism in small children is often overlooked and considered to be "growing pains." Billiet was opposed to the views of Roger, Thomson, Hughes and Bright, because the coincidence of chorea with rheumatism is far below the frequency of rheumatism as a disease, and furthermore chorea affects females more frequently than males, while the opposite is true of rheumatism. Romberg could see no relation between chorea and rheumatism. Wunderlich believed the pains, so often manifested in chorea, to be an expression of hypersensitiveness and not of rheumatism. Steiner (1868) saw only 4 cases of articular rheumatism in 252 cases of chorea.

Gowers found a history of rheumatism in 25 per cent. of his cases.

Osler found in 15.8 per cent. of his cases of chorea definite articular swellings, and in 5 per cent. of them rheumatic pains; 26 per cent. of Starr's cases gave a rheumatic history. Heubner describes chorea as an infectious disease, and regards it as much an evidence of rheumatism as gumma is of syphilis. Tylden reports in the St. Bartholomew's Hospital Reports a rheumatic family of personal history in 72 per cent. of his cases. In the 439 cases of the British Chorea Committee, 97 cases—or about 22 per cent.—gave a history of rheumatism. R. S. Eustis found among 60 cases that 9—or 15 per cent.—were rheumatic on admission, and 19 more gave a history of previous rheumatism in the muscles and joints, making a total of 28 cases, of which 46.6 per cent. could be classed as rheumatic, and 13 cases, or 21.6 per cent., gave a history of both rheumatism and chorea. In the writer's last 30 consecutive cases, 11 gave a rheumatic history.

Macalister,⁵ in studying the life of the leukocyte, has shown that while the toxin in the blood plasma of chorea is toxic to the leukocytes of healthy persons, the blood plasma in cases of rheumatism is scarcely at all toxic, and that the plasma from chorea cases was toxic to the leukocytes of rheumatic cases; some authors conclude from this that the poisons in the two diseases are dissimilar. In spite of this, a study of the statistics of this subject justifies the conclusion that rheumatism bears a close relationship to chorea in about 25 per cent. of the cases.

Much speculation has been indulged in as to the nature of these relations. Some observers believe that emboli pass from the heart to the brain, and by occluding the small vessels, produce softenings which give rise to the choreiform movements. This cannot hold true, because, as will be pointed out under the pathology of chorea, emboli have only been found in a small number of cases, and then again, rheumatism may give rise to chorea without producing endocarditis. Others think that the infective process may cause thromboses in the small cerebral vessels. Pathological studies do not support this hypothesis.

With the advances made in recent years in bacteriological research, the attempt has been made to ascribe chorea as due to *bacterial invasion*.

Wassermann described a streptococcus, which he believed to be the cause of chorea. When injected into a guinea-pig it caused a rise in temperature, swollen joints and choreiform movements. Dana found a coccus in a case of chorea with leptomeningitis of the brain and upper part of the spinal cord. Sachs has found a streptococcus in the blood of one of his patients with chorea. Meyer, Sander, and Cramer-Tobben also found streptococci in chorea. In a severe case of chorea and endocarditis, Triboulet found a bacillus. In two cases, Apert found Triboulet's coccus; Westphal found a staphylococcus in the blood and brain of a patient with severe chorea following articular rheumatism, and which, when introduced into the blood of animals, produced articular rheumatism.

Poynton and Paine cultivated the *Micrococcus rheumaticus* in cases of articular rheumatism, and they found the same organism in the brain in cases of chorea, associated with rheumatism. Beaton and Walker found the same coccus in several cases of chorea. Camisa and Guervier have independently reported the finding of cocci similar to those of Poynton and Paine. Collins, in 1914, reported a case of chorea cured by the injection of an autogenous vaccine prepared from a coccus obtained by lumbar puncture.

Richards, of New York, found in the blood of two cases of chorea the *Streptococcus viridans*—an organism culturally similar to the green-producing streptococcus of Schottmueller, and to that found by Poynton and Paine. Both Hastings and Thro corroborated the identity of Richards' streptococcus with that of Schottmueller. It may be noted here, however, that in these 2 cases the cerebrospinal fluid was cultured and no microorganisms were found.

Dick and Rothstein⁶ isolated a streptococcus from the throat of a patient who had been suffering from chorea for 5 years, and injected it into a dog, which developed, 12 hours after the injection, typical choreiform movements.

H. W. Frink⁷ reports the case of a girl of 15 with chorea, from whose tonsils Thro isolated a pure growth of the *Streptococcus viridans*, similar to the *Micrococcus rheumaticus* of Beattie, an autogenous vaccine of which cured the girl's chorea.

Koplik⁸ cultured the blood in many cases of chorea, but has never been able to find any microorganisms. Israel Strauss,⁹ in 1915, cultured aerobically and non-aerobically the blood and spinal fluids of seven cases of chorea, but the *Streptococcus viridans* could not be demonstrated. He also inoculated monkeys intracranially with the spinal fluid of all the cases used. In one case, this had resulted in typical chorea for twenty-four hours, which then disappeared and returned for shorter periods. The monkey was killed and the brain was found normal; the heart muscle was examined for Aschoff bodies, which were looked upon as pathognomonic of rheumatism, or a disease, like rheumatism, in which an organism had not been obtained, but none were found. There had been two cases of chorea in which the heart showed Aschoff bodies. The individual, from whose spinal fluid the monkey developed chorea, died, and the autopsy showed encephalitis with hemiplegia. The cortex was removed through aspiration, and there had been hemorrhage. The material was then inoculated into monkeys but nothing developed from it. In the other cases the same procedure was repeated and nothing was found.

J. Donath¹⁰ made bacteriological examinations of the blood, cerebrospinal fluid and brain tissue in 7 cases of severe chorea. Two of the patients had amentia as a complication; 2 had chorea gravis which terminated fatally, and 2 of the uncomplicated cases had such severe twitchings that the blood had to be taken under narcosis. In 5 of the cases he found the *Staphylococcus albus*, and in 4 of these 5 cases the germ was isolated from the blood, and in the other, from the brain tissue. In the remaining 2 cases the *Staphylococcus pyogenes aureus* was obtained from the blood and spinal fluid, respectively. In one case, *Sarcina lutea* was found in the brain tissue, and in another, in the blood. In some of the cases undifferentiated diplococci were also obtained from the blood. This author has no doubt of the infectious nature of the disease, but he does not think that these bacteria are the specific cause, but that they predispose feeble and anemic individuals to the disease.

The clinical fact that tonsillitis, abscessed teeth, aural discharges, and other focal infections have been followed by chorea which was promptly cured after the removal of these foci, is considered as an argument in favor of the infectious nature of the disease. In a study of 1,000 tonsillectomies performed by Crowe, Watkins and Rothholz, they found that

the removal of tonsils and adenoids is not a very satisfactory therapeutic or prophylactic measure in chorea; of the 23 cases of Sydenham's chorea, in which the tonsils and adenoids were removed by them, 8 had a recurrence of the chorea.

Helmholtz¹¹ found, in 138 cases of chorea, 33 per cent. with a history of repeated tonsillitis, 21 per cent. of rheumatism, 26 per cent. of endocarditis, 8 per cent. of rheumatism and tonsillitis, 21 per cent. of rheumatism and endocarditis, and 54 per cent. of the entire group showed manifestations of one or the other of these diseases. Looking at the problem from Rosenow's point of view, Helmholtz believes that the same organism which has a specific tendency to localize in the valves of the heart or in the joints may, under slightly different circumstances, localize in the brain cortex and produce chorea.

Quigley¹² cultured the tonsils, blood and spinal fluid in 21 cases of acute, subacute and chronic chorea; 11 patients gave a history of acute tonsillitis; 2 of rheumatism, and one of endocarditis; 8 gave no history of either of these, and none had any evidence of lues. The blood was cultured in all the cases and the results were positive in 10 of them—9 of these gave small, slightly elongated cocci arranged in pairs, short chains and groups, and the tenth positive blood-culture yielded a Gram-positive short diphtheroid organism. The spinal fluid of all the patients was cultured with thirteen positive results. Of these, 8 were organisms resembling those found in the blood. The throats of 15 of the patients were cultured, and 29 cultures were isolated and studied. The results showed: 10 were hemolytic streptococci of variable size, occurring in long or short chains; 13 were green-producing cocci growing in pairs and short chains, and 6 were organisms which grew on blood agar as pinpoint, colorless, non-hemolytic colonies resembling the organisms from the blood in their morphologic and cultural characteristics.

Kinsella and Swift¹³ undertook to determine whether any constant cultural or immunologic type of bacterium was associated with acute rheumatic fever. They studied 58 cases and their conclusions are: (1) that no type of streptococcus is constantly associated with acute rheumatic fever; (2) that the etiological relationship between the streptococcus and acute articular rheumatism cannot be definitely proven; (3) that, if the streptococcus is the etiological agent in acute rheumatic fever, it is through the various members of the *viridans* group, and hence no one member can be called the *Streptococcus rheumaticus*.

W. Lintz,¹⁴ from his researches on rheumatism, concludes that in some cases a microorganism can be isolated from the blood, but that the reason it cannot be found more frequently is because the bacteria tend to localize in the Aschoff nodules, and, except in the very virulent forms of the disease, are rapidly destroyed in the circulation. This investigator is not positive as to the exact nature of the organism, but he believes it to be similar to a streptococcus, and the reason why some animals fail to contract the disease after inoculation may be due to the lack of susceptibility on the part of the animal, or to the attenuation of the microorganism, or to both of these factors.

Morse and Floyd,¹⁵ from a study of 26 cases of chorea, are also of the opinion that a microorganism, or group of microorganisms, may be the cause of the disease, and that the source of infection is probably in the tonsils or teeth.

Reinhold reports a case of fatal chorea following sinus thrombosis, complicating pregnancy in a girl of 20. Stern collected in the literature 26 cases of chorea following sinus thrombosis; they were all fatal, but the results of the pathological examinations do not harmonize. Unless it was the same infection to which the thrombosis was due which produced the chorea, it is difficult to trace the relationship between the two conditions.

Rosenow¹⁶ as a result of his experiments on dogs believes that chorea is due to a streptococcus having specific neurotropic and immunologic properties, and obtainable from the tonsils, nasopharynx and teeth of the affected persons. He found lesions in the heart valves of inoculated dogs resembling those found in chorea in man, and anatomic changes consisting of inflammatory areas within or adjacent to the motor centers or motor paths of the cerebrum, mid-brain and cerebellum.

Cases of cerebrospinal syphilis occasionally exhibit choreiform movements; these movements are rarely generalized; they are usually confined to one or more limbs or to the face depending on the site of the lesion, which in such cases is usually irritative in nature. This form of chorea is included among the "symptomatic choreas."

In recent years, by the general impetus given through modern biologic and therapeutic methods to the study of syphilis, an attempt has been made to attach undue importance to syphilis as an etiologic factor in chorea. A careful investigation of the statistics on this problem would seem to show that syphilis might be an accidental, but is not a direct etiologic factor in chorea. A child affected with lues might contract chorea just as readily as any other child; the same holds true whether the lues is acquired or hereditary. Nevertheless, when a case of chorea has lasted over three or four months, and does not seem to progress favorably under routine treatment, it is well to bear in mind that lues might be at the basis of the disease.

The influence of *infectious fevers* on the development of chorea is important. Chorea may develop after scarlet fever at periods varying from six weeks to six months after the attack of scarlet has subsided. Priestley found 13 cases of chorea following 5,355 cases of scarlet fever—a proportion of 1 in 412. In Carslaw's 533 cases of scarlet, only 3 were followed by chorea. In the British Chorea Committee's report scarlet fever is given as the sole antecedent cause of chorea in 6 per cent. of all the cases. Osler states that scarlet fever with arthritic manifestations may be a direct antecedent of chorea; about 25 per cent. of his cases of chorea gave a previous history of scarlet.

Sturges (cited by Osler) states that a history of previous *whooping cough* occurs more frequently in choreic than in other children. Osler's Infirmary records do not bear out Sturges' findings.

Measles as an antecedent to chorea is recorded in the British Chorea Committee's report in 26 per cent. of the cases, and in 7 per cent., it was the sole antecedent illness. Chorea may follow influenza, tuberculosis, typhoid, gonorrhea, small-pox, chickenpox, diphtheria, cerebrospinal meningitis, pyemia, bacterial endocarditis or any infectious disease, especially when accompanied with hyperpyrexia and severe constitutional disturbances.

Kinnicutt, Heinemann and others have pointed out a relationship between chorea and certain forms of malaria. In southern climates,

where the cerebral forms of estivo-autumnal malaria are common, disorders of motility varying from slight muscular twitchings to choreiform and even convulsive twitchings are frequent, but these never seem to appear in the first paroxysm. It is doubtful, however, whether these cases can be considered genuine choreas.

Symptomatology.—**CLINICAL HISTORY.**—In most of the cases the temperature remains normal throughout the disease. In severe cases, especially those due to or following infections, it may be 100°–102° F. (37.8°–38.9° C.). Uncomplicated fatal cases have been recorded with a hyperpyrexia (106°–108° F.—41°–42.2° C.). Asthenia, loss of weight and exhaustion due to insomnia and inability to take sufficient food are common in the severe cases.

PHYSICAL FINDINGS.—*Choreic Movements.*—The most characteristic symptoms of the disease are the rapid, coarse, involuntary, spontaneous, irregular and purposeless movements. They cannot be arrested for any length of time, and are increased by attracting the patient's attention, by excitement and by an effort to restrain them, or to carry out any volitional movement; they are ordinarily diminished during physical and mental rest, and cease during natural or induced sleep.

At the beginning of the disease only occasional twitchings of the face or hand are noticeable, but as the disease progresses they become more frequent and more marked, and finally continuous. They may become so violent that the muscles of the entire body appear as if in constant motion.

Owing to the abruptness, rapidity and irregularity of the movements, work requiring exact coördination, such as writing, sewing, playing the piano or walking, cannot be performed; coördinated movements of the fingers, such as apposition of the thumb and index or little finger are usually impossible.

Oppenheim has drawn attention to the tendency that these patients have—in addition to the irregular and uncontrollable choreiform movements—to purposeless associated movements. Foerster has shown that in chorea the orderly coöperation of the agonists with the antagonists during voluntary movement is lost, but that single muscles or muscle groups participate in the contraction, e.g., in extension of the finger the extensor communis digitorum acts without the interossei; in closing the fist the wrist is not extended, etc. There is, in other words, a typical ataxia. A choreic patient, on account of this ataxia, never performs the same movements twice in the same manner.

Weakness of Muscle Power.—The next most common symptom is weakness of muscle power. This weakness may bear no relation to the severity of the choreic movements, and it may be considerable in extent before the latter are recognized, but there is never anything like a complete paralysis. This muscle weakness is called "choreic pseudoparesis," and in many cases it overshadows and inhibits the movements. Close observation will show that the little patient uses one limb less than the other, or perhaps ceases to use it at all. He can still move the limb when ordered to do so, but he does so only for a short time and very feebly. The muscles become limp and hypotonic; the tendon reflexes are diminished, and in some cases even absent. As the disease advances the twitchings become more marked, and the pseudoparesis assumes secondary importance as a symptom. West called this condition "chorea mollis," and Gowers¹⁷ "chorea paralytica."

While any muscle of the body may be affected by the choreiform movements, they are more noticeable in the extremities and face than in the muscles of the trunk. The involvement may from the outset be general, but as a rule is confined to one limb, or to one side of the body, but becomes generalized as the disease is progressing. In at least half the cases both sides of the body are not equally involved. The arms are almost always affected earlier and more severely than the legs; the facial muscles are very frequently affected, resulting in peculiar facial contortions and grimacing; the eyes are suddenly closed and opened, the mouth pouts, the tongue rolls within the mouth and is pressed between the cheeks so that speech becomes quick and indistinct. The words are shot out or jerked out in separate parts, the last syllables being entirely cut off. Laryngoscopic examination discloses an irregularity in the movements of the vocal cords. Involvement of the diaphragm results in irregular and "catchy" respiration, and further interferes with speech. According to Glogau,¹⁸ the curve taken from thoracic breathing during speech will show choreic jerks, which cannot be demonstrated in other muscles.

Swift¹⁹ of Boston, from a study of the voice in 20 cases of chorea, has found that there is a change of voice which is more frequent in the vowels, less so in whisper, whistle, consonants, air-blow and holding of breath—the frequency being in the order cited. He found sufficient uniformity and frequency in the appearance of vocal changes to warrant a classification of changes in pitch and intensity of the voice, as one of the signs of chorea. The most marked change occurred in the open, prolonged sound of "a" as in "are." When the pseudoparesis involves the muscles of speech and respiration, the voice may be reduced to a whisper, and in severe cases the patient may become temporarily mute.

The involvement of the *tongue and mouth* may also extend to the pharynx and seriously interfere with the taking of food.

Involvement of the *muscles of the neck* causes frequent movements of the head to one side, the eyes moving with the head. The hands are alternately flexed and extended, the fingers are spread apart, the arms rotated inwards, and the shoulders are drawn up. Slight affection of the *trunk muscles* causes the patient to sway from side to side when sitting or standing, but a more severe involvement results in a total inability to sit or stand. The limbs may be thrown about so violently that the patient cannot rest in bed, and frequently sustains severe contusions all over the body. Tuckwell records a case where spasm of the *muscles of the jaw* was so violent that several teeth were broken. *Involvement of the legs* results in their being thrown about violently, now extended, now pressed against each other, then again rotated outward, or abducted.

As a result of unequal spasm of the muscles of the *eyeballs*, temporary diplopia may be present. The pupils are dilated but react to light and accommodation. Von Ziemssen observed on several occasions the pupillary reactions to be sluggish. Hippus (spontaneous, rapid and spasmodic variation in the size of the pupil) has been noticed. Ophthalmoscopic examination shows the fundi to be normal. Embolism, atrophy of the disk and optic neuritis have been seen during or after attacks of chorea, but these changes are not characteristic of the disease. Concentric restriction of the visual fields has also been noted in chorea.

Electrical Irritability of the Muscles.—Uncomplicated cases of chorea do not show any alteration in the electrical irritability of the muscles. These retain their size, but in "paralytic" cases they are hypotonic. Elloy claims to have seen muscular atrophy in the involved muscles.

Reflexes.—The tendon reflexes are, as a rule, normal; they may, however, be increased or diminished. Joffroy has seen cases where they were completely abolished. Oddo believes the reflexes are normal in mild cases, and diminished or suppressed in severe cases. In the writer's experience the reflexes have either been normal or increased, except in paralytic cases, when they were diminished, but never absent.

Gordon and Eshner have observed cases in which, when the patient is on his back and a short blow is struck over the patellar tendon, the response will be like in the ordinary knee jerk, but instead of the leg coming down immediately after, it will remain suspended in the air for some time and come down gradually, i.e., the knee jerk has the character of a tonic muscular contraction. Oppenheim considers this phenomenon to be a coincidence, or that it may possibly be due to a reflex choreic contraction of the quadriceps tendon occurring simultaneously with the reflex movement. This may also be an explanation of the "wobble knee-jerk," described by Swift of Boston as characteristic of chorea.

Sphincters.—The sphincters are not involved, except in the terminal stages of the very severe cases, and in the cases with marked mental symptoms, as a result not of paralysis but of mental apathy. Enuresis in children, however, is quite common both at the beginning and throughout the disease.

Sensation.—Chorea is a painless disease. The incessant movements cause fatigue but no actual pain. When the disease is due to or complicated with rheumatism, there may be pains in the muscles, joints and nerves. In uncomplicated cases there are no objective sensory disturbances.

Tremor.—A fine, regular tremor involving the tongue, lips and fingers may be observed in addition to the choreiform movements. This tremor is intensified after physical and mental excitement, and is probably due to the accompanying general debility.

Convulsions.—Convulsive attacks do not occur in chorea, except as manifestations of an associated hysteria or epilepsy. Gowers states that he has found in many cases of epilepsy that the convulsions made their first appearance after an attack of chorea.

Heart.—The most common symptoms referable to the heart are those due to mitral insufficiency. The organic nature of the murmur in this condition must be differentiated from the usual systolic murmur heard over the mitral or pulmonic areas, due to secondary anemia or conditions other than endocarditis. The latter is often associated with a venous hum in the jugular vein, and the murmur itself is not transmitted. The cardiac dullness, which may extend slightly to the right, may also be due to the anemia, and subsequently disappear. The endocarditis associated with chorea is usually of a mild nature, and may disappear without leaving any trace. Occasionally a faint systolic murmur may be heard at the apex with some beats and not with others, said to be due to irregular contraction of the papillary muscle. Cases complicated with rheumatism may show a friction rub over the precordial area due to a complicating pericarditis.

Pulse.—The pulse is usually increased in frequency about 10–15 beats per minute, and on account of the irregular breathing the heart's action may also be irregular. Irregularity, however, is not as marked a feature in chorea as rapidity. The subjects of the disease being usually neurotic children, slight emotional disturbances are sufficient to produce a rapid, irregular and tumultuous heart action.

Blood-Pressure.—Owing to the accompanying anemia there is a tendency to a somewhat lower blood-pressure than normal.

Skin.—Vasomotor disturbances, such as hyperidrosis, dermatographia, flushing of the face and neck, and general erythema, are seen in children with a tendency to spasmophilia and to ductless gland disturbances. Fissured lips, with herpes labialis due to irritation by the continuous smacking of the lips, are perhaps the most common symptoms referable to the skin and mucous membranes. Other cutaneous disturbances when present are probably due to poisoning with arsenic taken for therapeutic purposes. These are characterized by the presence of papular or erythematous rashes combined with pigmentation of the skin. Osler found two cases of herpes zoster in his series at the Philadelphia Infirmary. The various eruptions of purpura are not uncommon in "rheumatic" cases. English clinicians have described subcutaneous fibrous nodules in chorea, but no such cases have been reported in this country. Children, with the so-called exudative diathesis, are predisposed to chorea, and, inasmuch as such children are also frequently affected with eczema, the latter has been considered a complication or manifestation of the disease. Some writers have even gone so far as to claim that a child suffering from eczema is most apt to become choreic when the eruption of the eczema has been allowed to "strike in." It is doubtful whether any of the cutaneous manifestations seen in chorea are more than mere coincidences.

Rheumatism.—Rheumatism is so commonly seen in chorea that some writers consider it a symptom of the disease. In our opinion, it is a cause and complication of chorea, and is discussed in the sections on etiology and complications.

PSYCHIC STATES.—Mental symptoms play an important rôle in the symptomatology of chorea. Hammes²⁰ noted definite symptoms of a psychosis in 18 patients out of a series of 88 cases of Sydenham's chorea. As a rule the patients are irritable, peevish, and forgetful; they are easily frightened, cannot concentrate, are disobedient, and very fretful. Some of them have night terrors and transitory auditory and visual hallucinations. Psychic symptoms other than irritability, fretfulness and disobedience are not as common in children as in adults. In adults the psychic symptoms are not unlike those of the infectious or toxic deliria. The intellect, as a rule, does not suffer. The mental symptoms are at their maximum at the height of the disease, and may persist after recovery. Severe mental symptoms make the prognosis much graver. Cases of chorea have been recorded in which the movements were insignificant as compared with the psychic disturbances.

LABORATORY FINDINGS.—*Cerebrospinal Fluid.*—In ordinary cases the cerebrospinal fluid shows no deviation from the normal. In cases in which the bacterial origin of the disease can be traced to some localized or general infection, bacteria may be found on culturing the fluid.

Blood.—In the majority of cases the blood shows an anemia of the chlorotic type with little change in the number and character of the

erythrocytes but with a low color index. Leopold²¹ of Philadelphia found in 20 cases of chorea an eosinophilia varying from 4 to 16 per cent. F. H. Leavitt²² examined the blood in 80 cases of Sydenham's chorea in the active stage of the disease; he paid special attention to the frequency of eosinophilia and its relation to herpes labialis. He concludes: (1) That herpes labialis is most likely due to mechanical irritation by the patient himself, and that eosinophilia, when present, is due to the condition of the skin, and not to the chorea itself. (2) There is a great increase in the lymphocyte and a relative decrease in the polymorphonuclear count. (3) A fairly constant low-grade leukocytosis is present in most cases, indicating the infective origin of the disease.

Urine.—The urine is abundant in amount, and when the chorea is associated with rheumatism, large amounts of urates, uric acid and phosphates are in evidence. During the height of the disease the specific gravity may be 1.030 or even 1.035. Changes in the urea nitrogen output, albumin and casts, in the absence of cardiorenal complications, are very rare. Temporary glycosuria has been observed.

Duration.—The average duration of the disease is from six weeks to six months, but it may last only three weeks, or more than six months. The average duration of the cases collected by the British Chorea Committee was about ten weeks. As a rule, the severer the disease the longer is its duration. The age and sex of the patient, the condition of the heart or the other complications do not seem to bear any relationship to the duration of the disease. Slight choreiform movements, the so-called "residual chorea" (Guthrie) may persist for months.

Recurrences.—Recurrences in chorea are so common that they are considered a characteristic feature of the disease. Two or more recurrences are quite common. Sachs and Peterson found that, out of 70 cases, 18 had a second attack, 11 had a third, 4 had a fourth, 1 had a fifth, and 1 had a sixth attack. Females are more liable to recurrences than males. The disease seldom recurs between the ages of 18 and 30, except in pregnancy. After the disease has once ceased, the slightest cause such as fright, overstrain, or an acute illness, may bring on a recurrence. Endocarditis and pericarditis seem to develop more commonly during a recurrence than during a first attack.

The intervals between the relapses may vary from a few weeks to one or two years; they may vary in the same patient, and there is no uniformity as to the time of the year in which a relapse may occur, as compared with the previous attacks. A recurrence may imitate the first attack as regards its mode of origin and site of development of the choreiform movements, but there is no definite rule about this, as there is none about the severity of the relapse as compared with the severity of any of the previous attacks.

Diagnosis.—The disease is easily recognized by the peculiar movements which can be elicited in the mildest case by making the patient hold both hands extended above his head; after a few seconds, especially when his attention is distracted, rapid, irregular twitchings appear in the fingers. Another good method of eliciting the movements—particularly in the mild, unilateral cases—is to let the patient grasp the examiner's hands; the difference between the uniform muscular contractions of the healthy side and the unsteadiness of the grasp on the affected side will then be readily noticed. In the cases in which the

legs are more involved than the arms, the station and gait may simulate paraparesis, but the twitchings will be sufficiently evident to show the exact nature of the trouble.

DIFFERENTIAL DIAGNOSIS.—*Paralytic Chorea*.—*Unilateral Multiple Sclerosis*.—In paralytic chorea, when loss of muscle power is a predominating feature, diagnostic difficulties may arise. Such cases may be mistaken for genuine paralyzes. In "paralytic chorea" the weakness is, in most cases, confined to one arm; it does not involve the face or leg, and on careful examination choreic movements will be observed; these, with the history of the gradual loss of the use of the affected extremity, will clear up the diagnosis. Grinker of Chicago, showed a case before the Chicago Neurological Society on October 21, 1909, which he had seen three years before that and had made the diagnosis of paralytic chorea; at that time the patient began to show clumsiness on one side of the body, and later, irregular choreiform movements began to make their appearance on the same side; there were no other signs. As time went on the patient developed a scanning speech with all the other classical signs of a unilateral multiple sclerosis.

Tic.—In distinguishing chorea from tic it must be borne in mind that in tic the movements are rapid, coördinate and purposive; they may be tonic or clonic, with comparatively long periods of rest between the movements. Tic is usually confined to certain definite parts of the body, most commonly the face and shoulders, rarely to the trunk or legs. A choreic does not repeat the same movement as regularly and as systematically as a tiqueur. No matter how violent a tic may be, it never interferes with voluntary movement. Tiqueurs are usually psychically abnormal.

Spasm.—Chorea is differentiated from spasm by the fact that in the latter the movements are very brusque and involve muscles corresponding to the anatomic distribution of a certain nerve. They are usually due to some reflex irritation along the reflex arc in that distribution. Examples of such spasms are blepharospasm, facial spasm, torticollis, etc. A spasm may begin in a single muscle and spread to neighboring muscles. In spasm, the muscles may become hypertrophied from overuse, but this condition never occurs in chorea. (See chapters on Spasm and Tic.) When the chorea is associated with tic or spasm, the diagnosis may be very difficult.

Myoclonias.—Chorea is distinguished from the myoclonias by the fact that in the latter the twitchings occur in paroxysms, and are of lightning-like rapidity (30–120 per minute), clonic in nature, and involve a part of a muscle, a single muscle or a group of muscles, and rarely, if ever, produce movement of the parts involved. They are usually associated with other nervous diseases, especially epilepsy.

Choreo-athetoid Form of Infantile Cerebral Palsy.—Chorea, which has existed from early childhood, may be mistaken for the *choreo-athetoid form of infantile cerebral palsy*. The muscular rigidity, especially of the legs, with other signs of pyramidal tract involvement, the athetoid character of the movements, and the fact that congenital or early acquired infectious chorea is very rare will prevent confusion.

Athetoid Movements.—Choreiform movements differ from athetoid movements by the fact that the latter occur in spastic, incompletely paralysed limbs, and that they are slow, rhythmic, twisting and "jelly-fish".

like in character involving the distal ends of a limb, and that the face is rarely involved, except in athétose doublé.

Fibrillary Twitchings—Myokymia.—The fact that these movements are confined to muscle bundles rather than to entire muscles and that they never give rise to a movement of a segment of a limb or of an entire limb is sufficient to differentiate them from choreiform movements.

Tetany.—The spasms in tetany are characterized by being painful and by involving bilaterally the hands and feet, giving rise to characteristic postures ("main d'accoucheur," etc.). The presence of Chvostek's, Erb's and Trousseau's signs (see Tetany) will be diagnostic.

Tetanus.—The characteristic tonic spasms of the jaws and face, trunk and limbs, with complete relaxation between the spasms, and the history of the mode of onset will readily differentiate tetanus from chorea.

Hysteria.—The age of the patient, the absence of a cardiac or rheumatic history, and the presence of the usual stigmata of hysteria, and of a peculiar mental make-up, with the history of a sudden onset of polymorphous muscular twitchings in isolated parts of the body, and their frequently rhythmic character, after the hysterical patient has had an opportunity to observe a patient with genuine chorea, will aid in the differentiation of the two conditions.

Dystonia Musculorum Deformans—Wilson's Disease—Westphal-Strümpell's Pseudosclerosis.—These conditions are also characterized by the presence of irregular involuntary spasmodic twitchings of the muscles, which may on superficial observation simulate choreiform movements. The clinical course of these diseases, however, is so different from that of chorea that ordinarily these conditions can readily be distinguished from Sydenham's chorea.

Huntington's Chorea.—Sydenham's chorea is differentiated from Huntington's chorea in that the latter, as a rule, begins after the age of thirty, is both hereditary and familial in nature, progressive in its course, with gradual mental deterioration leading to complete dementia. In the absence of a history of heredity, a case of chronic Sydenham's chorea will require long observation to make the differentiation between the two conditions positive.

Complications.—Endocarditis.—Endocarditis is frequently met with in individuals who have or have had chorea. Some authors consider endocarditis a symptom, others an etiologic factor in conjunction with rheumatism, and still others a complication.

Sturges collected 80 fatal cases and in only 5 of them was the heart normal. Osler found in 170, out of his 554 cases of chorea, heart murmurs—149 of these were at the base. Of the 449 cases reported by the Committee on Collective Investigation of the British Medical Association, 113 had cardiac murmurs; how many of these were organic and how many were functional could not be ascertained. Stephen MacKenzie examined 33 patients varying from 1 to 35 years after an attack of chorea and noted signs of undoubted cardiac disease in 60.6 per cent. Osler, out of 140 cases (post-choreic) found the heart normal in 51 cases; in 17 there was some cardiac disturbance which could be considered functional, and in 72 cases or 51 3/7 per cent. there were definite signs of organic heart disease. Fagg found heart disease in 17 out of 18 cases of chorea at necropsy; in 5 of these death was not due to the severity of the chorea. R. S. Hustis,* of 60 cases studied in the Children's Heart Clinic at the

Massachusetts General Hospital found 32 or 53.3 per cent. to be choreic on admission, and 10 more gave a previous history of chorea, making a total of 42 cases or 70 per cent. in which chorea played a part. Abt and Levinson²² found 73 cases of heart disease in 226 cases of chorea, the majority of which were diagnosed as mitral insufficiency, one as myocarditis, two as single aortic insufficiency, and one as double aortic.

In the majority of cases of heart disease complicating chorea the lesion is one of mitral insufficiency. The murmurs in these patients are due to structural changes in the mitral valve produced by the poison of rheumatism, which, as has been pointed out under etiology, plays such an important rôle in the causation of chorea. The finding of this form of cardiac disease in the cases of chorea not due to rheumatism or any other infectious agent is very difficult to explain. A. Ernest Sansom²⁴ considers the signs and symptoms of mitral insufficiency in the non-rheumatic cases different from those which are due to rheumatism. In some of these, he claims, careful examination for many days may fail to elicit any evidence of valvular involvement, but later a soft systolic murmur becomes audible at the apex; there is no accentuation of the second pulmonic sound; the ventricles do not become dilated and yet the murmur retaining its original character persists for several years becoming completely inaudible later in life. Pathological evidence completes the distinction. In these non-rheumatic cases, according to the same author, the left auriculoventricular orifice on its auricular aspect has been found studded and fringed with small papilliform elevations of the endocardium; these are firm to the touch and cannot be detached by rubbing with the finger. The endocardium covering these elevations remains perfectly smooth. They do not begin, as in rheumatic endocarditis, with a change in the epithelium and an attachment to the roughened surface of the fibrous caps, but they are firm outgrowths showing fibrous hyperplasia. This type of endocarditis is not followed by sclerous changes with retraction of the valves, cords and columns, and the endocardium remaining smooth; there is very little interference with the closure of the valve in systole. These changes, says Sansom, "may be the immediate results of a sudden overstrain and rupture of the terminal arterioles distributed to the valve structures." Experimental production of overstrain and fright in animals by Roy and Adami were followed by similar changes in the endocardium.

Pericarditis.—Pericarditis is occasionally met with as a complication of chorea, especially in the rheumatic cases. It is not nearly as common as endocarditis, and presents no unusual features.

Rheumatism.—The rôle which rheumatism plays in the causation of chorea has been discussed under the etiology of the disease. Its association with chorea may manifest itself in three ways: (1) an attack of articular rheumatism will precede by months or years the onset of an attack of chorea, and will not recur before or during the entire course of it; (2) an attack of articular rheumatism will begin simultaneously with the choreiform movements; (3) it will appear during the course of chorea.

Articular rheumatism during chorea is, as a rule, milder in its course than ordinarily. Patients with chorea frequently complain of vague pains in the joints, which are too readily ascribed as due to rheumatism. It is well to bear in mind that while chorea is not a painful disease, and is accompanied with no sensory changes, nevertheless, when the dis-

case is very severe, the constant wriggling and throwing about of the limbs may give rise to pains. Osler asks whether these pains are not analogous to those seen in the limbs and joints during most infectious diseases. The French writers call these pains, for which no local cause can be discovered, *choreic arthropathies*.

Psychoses.—Chorea is frequently complicated with various psychoses; this is more commonly so in adults than in children. In these cases it is important, both from a diagnostic and prognostic standpoint, to differentiate the existence of a psychosis "per se" from the ordinary psychic states which are seen in the disease itself. The mental symptoms may be the predominating feature and overshadow the chorea with the result, as Osler says, that patients have even been committed to the insane asylum.

Feeble-mindedness.—Congenital as well as acquired feeble-mindedness may be combined with chorea.

Epilepsy—Hysteria—Spasms—Tics.—The association of chorea with epilepsy is not as common as with hysteria and the other neuroses such as spasms and tics.

Cerebral Diplegias and Hemiplegias—Athetosis and Other Organic Nervous Diseases.—Chorea may also complicate the cerebral diplegias, the hemiplegias and the various forms of athetoses and other organic nervous diseases.

Exophthalmic Goiter.—Choreiform movements may occur in exophthalmic goiter, and the two conditions may be combined.

Anemia.—The association of chorea with the chlorotic type of anemia is so common that anemia may be said to be one of the cardinal symptoms of the disease rather than a complication.

Jaundice.—Jaundice, according to Poynton, is very common in rheumatic children, and it may complicate the course of a severe chorea.

Chorea in Pregnancy.—General Considerations.—The influence which pregnancy exerts on the occurrence of chorea is still unknown. It is commonly regarded as a reflex irritation similar to that which causes the vomiting of pregnancy, but inasmuch as the development of chorea in pregnancy is usually later than that of morning sickness and rarely ceases immediately after the removal of the products of conception by labor or abortion, the analogy seems hardly tenable. There seems to be no doubt, however, that the fetal movements are the actual exciting cause.

Symptomatology.—The choreic movements may manifest themselves at any time during pregnancy; they are, however, most common in the third or fourth month, and rarely begin in the eighth or ninth month.

The nature of the movements, their onset and involvement are similar to chorea in the non-pregnant. The movements may extend to the uterus. Braxton Hicks²⁵ reports the case of a young woman who had chorea in childhood. During pregnancy the chorea had recurred, and the uterus, which could be distinctly outlined in the abdomen, presented a marked alteration in form, accompanied by evident choreic contractions. Rest in bed with arsenic diminished the movements. Labor was uneventful and she made a good recovery.

Romberg was the first to point out that chorea in pregnant women is more apt to be bilateral and to involve the tongue.

The psychic manifestations of chorea in pregnancy are characterized

by greater memory defects and maniacal outbursts than in the non-pregnant. The memory, however, improves with the cessation of the chorea. Maniacal choreics give peculiar outcries not unlike patients about to be seized with an epileptic convulsion. The irritability, the visual and auditory hallucinations, the dream-like confusion with the peculiar lack of connection of ideas do not differ from those seen in the non-pregnant, except that their prognosis is much more unfavorable because they have a tendency to persist after the chorea has ceased.

After delivery the choreic movements gradually subside, but they have been known to continue for 4 or 5 months after labor or abortion and in a few cases to have remained permanently.

Occurrence.—**RECURRENCES.**—Pregnancy always aggravates an existing chorea and predisposes to recurrences. Acute rheumatism is generally the most immediate cause. Fright, emotional disturbances, infection and anemia favor its occurrence. As in the non-pregnant, a recurrence during pregnancy may be more severe than a former attack. Primigravidæ are more susceptible than multigravidæ. The younger the patient the greater the tendency to recurrences. Chorea of pregnancy rarely occurs for the first time after the age of twenty-five in a patient free from rheumatism.

If chorea recurs during several pregnancies there is no uniformity as to the time of its onset. Lawson Tait observed a patient who was rheumatic and had chorea at sixteen; she had a recurrence in the fourth month of her first pregnancy, in the third month of the second, at the beginning of the third, and in the fourth month of the fourth pregnancy. There are cases on record in which a first attack of ordinary chorea began after delivery or after abortion.

Effect of Chorea on Pregnancy and Prognosis.—This depends upon the severity of the disease. In mild cases, if properly treated, the pregnancy may not be interrupted. In severe cases abortion may occur spontaneously. When this occurs the prognosis is very grave. Barnes²⁶ collected 56 such cases with a fatal termination in 1 out of every 3; most of the patients developed septicemia or pyemia which was followed by hyperpyrexia, coma and death. The earlier in pregnancy the chorea occurs the greater the danger for the child. The mortality of chorea in pregnancy is unusually high. The onset of labor during chorea adds to the seriousness of the prognosis. In a series of cases recently reviewed, 438 in number, which included all grades of the disease, the mortality in the mothers was 16.5 per cent. The mortality of the newly-born is also very high; many of them are still-born. It is interesting to note that some of the children which survive may show choreic symptoms shortly after birth, while others are perfectly normal.

Treatment.—The treatment of chorea in pregnancy does not differ from the treatment of ordinary chorea. The **salicylates**, **arsenic**, **rest**, and **hydrotherapeutic measures** are indicated. **Chloral**, **luminal**, and the **bromids** are useful in cases with intense restlessness and severe insomnia.

Obstetrical Indications.—In the severe cases, exhaustion, insomnia, marked mental symptoms or grave physical complications are indications for the artificial termination of pregnancy. It is well to bear in mind that these patients are predisposed to hemorrhages, both on account of the generally impoverished condition of the blood and the usual previous

administration of large doses of bromids for the chorea. Owing to the severity of the choreic movements during labor, the second and third stages may be unusually difficult. The patients may actually have to be held down by attendants, or anesthetized, to prevent them from seriously injuring themselves (Kolde²⁷).

The successful cure of pregnancy toxicosis by serotherapy has prompted some obstetricians to use this method in the treatment of chorea in pregnancy. Albrecht²⁸ reports a case in which a patient of his had chorea during her first pregnancy; it recurred during the second, and treatment with the ordinary methods for 22 days gave no relief; she was then injected with 20 c.c. of serum from a normal pregnant woman, and within 24 hours the chorea that had tormented her for over 3 months subsided permanently. He concludes his report by suggesting that a similar toxic action during the prepuberty stage may be the explanation of chorea in the young; the approach of puberty producing changes in the glands of internal secretion like the changes inaugurated by gestation.

An interesting case of chorea during pregnancy, bearing on the possible psychogenic origin of the disease, is reported by Flamma.²⁹ A woman of 21 developed an attack of chorea in the third month of her first pregnancy. The chorea growing worse an abortion was induced after which it gradually subsided. Becoming pregnant a year later the chorea recurred in the second month. The woman and her husband begged to have an abortion done, as the choreic movements were incessant and severe. Believing that the trouble was principally of nervous origin, a sham abortion under chloroform anesthesia was resorted to, after which the chorea disappeared. The pregnancy was uninterrupted, and she was delivered at full term. Flamma found in the literature many cases where no other factor than the pregnancy could be incriminated to account for the chorea. He remarks that pregnancy chorea and chorea in the pregnant are not necessarily the same thing. Sham interruption of the pregnancy proved effectual in another case in his clinic, in which the psychogenous manifestation took the form of uncontrollable vomiting. The two cases with the same origin but different manifestations, cured by the same measure, is believed by Flamma to prove the conception of a psychogenous origin in such cases. He advises before inducing actual abortion in dubious cases of this kind to attempt a sham obstetrical intervention.

Clinical Types.—MILD OR SIMPLE FORM.—In this form, which is the most common, the disease develops slowly and insidiously, rarely suddenly. The children become peevish, restless and irritable; they become awkward and clumsy and begin to drop things; in school they cannot sit quietly; they write poorly, pay no attention to their teachers, bite their finger-nails and pull the buttons off their clothes. They sleep poorly, have night terrors and enuresis. They shrug their shoulders, twitch their lips, wrinkle their foreheads and wink their eyes. Punishment has no effect; if anything, it aggravates the condition.

As time goes on they become pale and weak, their appetite poor and capricious, and digestive disturbances become frequent. Headaches and vague pains in the extremities now make their appearance. At this time the typical choreiform movements become more and more noticeable; at first in one arm, or one leg, or face and arm, or leg and arm, until even-

usually the musculature of the entire body is involved. These movements may last from three weeks to six months, depending on the severity of the disease. During this time there is usually a period during which the disease is at its height, after which there is a gradual regression of the symptoms. The movements now become less severe and less frequent; later they are observed only during emotional or physical excitement, and finally disappear altogether. The incoördination and weakness disappear before the movements have entirely ceased. In this form the spasms are slight and speech is almost never seriously affected.

SEVERE FORM.—The disease, as a rule, develops suddenly after fright, trauma or shock. The movements are general, and the involvement extensive at the very beginning of the disease. They are so violent that they interfere with speech and locomotion thereby incapacitating the patients from dressing and feeding themselves. In this form there may be loss of power in the extremity most involved, giving rise to so-called "paralytic chorea." These cases are usually of longer duration than the milder forms.

MALIGNANT CHOREA OR CHOREA INSANIENS.—This is the gravest but, fortunately, the rarest form. The cases belonging to this group may follow the simple or severe form, or they may be the terminal stages of either one of these, or they may begin as malignant forms. In the latter cases, it is most frequent in the second half of the second decade of life. This form is characterized by a predominance of the mental over the hyperkinetic symptoms. The patients become maniacal, delirious and confused; they have delusions and hallucinations; they cannot sleep, the temperature rises to 104° F. (40° C.) and sometimes as high as 106° F. (41.1° C.), and death from exhaustion is the usual termination. The mental symptoms are so marked that they are frequently mistaken as symptoms of an ordinary toxic psychosis. The simple delusional states which one meets at times in the other forms of chorea are insignificant in comparison with those seen in this grave form, and can hardly ever be confused with them.

Treatment.—**GENERAL MEASURES.**—Because the disease is one of long duration, it is important to keep the patient under the best general hygienic conditions possible.

HYGIENIC MEASURES.—Localized infectious foci should be removed, diseased tonsils excised, discharging ears treated, decayed teeth extracted, and general oral hygienic measures resorted to. The food should be abundant, light and nourishing. In cases complicated with rheumatism, red meats should be given sparingly. Plenty of water and fresh or stewed fruit are indicated. In cases with intestinal indigestion, the daily irrigation of the intestinal tract is advisable. Lactic acid cultures have been found to be of value in such cases. Tea, coffee and sweets should be used moderately.

Rest.—Absolute rest is essential, and in severe cases isolation with a tactful nurse indicated. Excessive mental or physical exercise is to be forbidden. Ambulatory cases are not to be allowed to attend theaters, or moving picture exhibitions.

MEDICINAL TREATMENT.—The three most important drugs in the treatment of chorea are arsenic, antipyrin and the salicylic acid group. Arsenic has been employed with success as far back as 1870 by Alexander, and recently by Pawlow, Bokay, Marie and others in the form

of ~~salvarsan~~. in the treatment with arsenic it is important to bear in mind the untoward effects of the drug. Arsenical neuritis, gastritis or nephritis may result from the injudicious use of the drug and may then be as serious as the chorea itself. The writer believes the administration of arsenic in the following way will minimize its possible untoward effects: For a child eight years old begin with 4-5 drops of Fowler's solution, well diluted, three times a day after each meal, to be increased daily by one drop till 12-15 drops have been taken three times a day. Seguin³⁰ regards 18-27 drops three times a day a normal dose. The parents are instructed to watch for nausea, vomiting, pain in the stomach, puffy eyelids and severe neuralgic pains in the extremities on the days when the larger doses are given. If such symptoms do occur, say on the sixth day, it is established that this particular patient's maximum dose is ten drops three times a day. After this, the patient receives no arsenic for three or four days; this gives him an opportunity to get rid of the untoward effects of the arsenic already ingested, after which the original dose of four drops, three times a day, is resumed and gradually increased till he obtains ten drops t.i.d.—the maximum dose, which he can apparently tolerate.

Eulenberg and Hammond have used arsenic hypodermatically. Bokay³¹ administered 0.2 gram (3 grains) salvarsan subcutaneously to a child 8 years old; improvement began on the second day; it was striking on the fifth, and after four weeks recovery was complete. Another child with a less severe chorea under treatment with Fowler's solution made a much slower recovery. Two cases with recovery in 15 days after the injection of 0.2 gram (3 grains) are reported by Hainiss,³² and Benno Hahn³³ reports three cases treated with intravenous injections of 0.08-0.3 gram (1.3-5 grains) and recovery in from 8 to 28 days. Each of Hahn's patients received three injections three to eighteen days apart.

We have used sodium cacodylate intramuscularly, and neosalvarsan intravenously, with no better results than with Fowler's solution by mouth. Arsenic may also be used by mouth in the form of arsenious acid (grains 1/130-1/60); its combination with iron in cases in which anemia is a marked feature, and with digitalis in cardiac cases, is advisable.

Antipyrin has always been a favorite remedy for chorea with foreign clinicians. It is best administered in doses of 5-8 grains (0.324-0.52 gram) three times a day. Comby always uses antipyrin in very large doses, 90-140 grains (5.85-9.10 grams) a day, and if the disease does not respond to these doses, he employs arsenic.

Salicylates are indicated when chorea is complicated with manifestations of rheumatism; they are best administered with alkalis to prevent gastric irritation. Langmead uses salicylates only when the disease is accompanied with very high temperature; he does not believe that this drug has any effect on the severity or duration of the chorea. Pearce Bailey³⁴ advises the use of aspirin, whether the cases are complicated with rheumatism or not, but he insists on the importance of absolute rest and isolation with the application of cold packs and occasionally lumbar puncture when the fluid is under high pressure. He advises, after the movements have ceased, a three or four weeks' stay in the hospital for the purpose of reestablishing the tone of the nervous system and thereby diminishing the tendency to a relapse. He very rarely had occasion to use ~~salvarsan~~.

For the motor restlessness, chloral, bromids, belladonna, hyoscyamus, hyoscyamin, hyoscin, apomorphin, gelsemium, cannabis indica, allonal, luminal, and even small doses of codein and morphin may have to be administered.

INTRASPINAL AND SUBCUTANEOUS INJECTIONS OF MAGNESIUM SULPHATE.—Following Meltzer and Auer's²⁵ experiments with magnesium sulphate used intravenously, subcutaneously and intraspinally for the production of regional anesthesia and for the treatment of tetanus, Marinesco employed this drug intraspinally in Sydenham's chorea with good results. Similar results were reported by Baduel, Rocaz, Calcaterra, Caronia and others. Augusto Natali²⁶ reports the successful treatment of eight severe cases of chorea by the so-called Marinesco method. His technic was as follows: He performed lumbar puncture in the usual manner, evacuated a certain amount of cerebrospinal fluid, and injected a solution of magnesium sulphate, the strength of the latter varying from 7 to 25 per cent. and the amount injected was 1 c.c. for every 25 pounds of body weight. All the cases had previously been subjected to the usual methods of treatment without any signs of improvement. After treatment with magnesium sulphate intraspinally, in 6 of the cases, the results were remarkable; in 2, one of which was a chronic case, there was also a marked improvement, but it was very slow. Natali points out the advantage of following the treatment by means of a brief course of arsenical medication; he does not believe that the magnesium sulphate acts as a specific, but is merely a symptomatic remedy diminishing the excitability of the nerve centers.

Excellent results have been reported by R. Pastore²⁷ in 4 cases of chorea in which magnesium sulphate was given intraspinally in small repeated doses. She gave 5 to 7 injections of 0.1–0.2 gram (1.5–3.0 grains) of magnesium sulphate in a 25 per cent. solution to a total of 0.4–0.5 gram (6.0–7.7 grains); the intervals were from one to two days at first and three to seven days later. There was no reaction; one child improved after the third injection; another child after the sixth, and another after the seventh. In one little girl of eleven, the third injection was followed by severe headaches, superficial breathing, sluggish pupils and a return of the choreic movements, but upon two more injections given after this, pronounced improvement set in.

Heiman,²⁸ of New York, treated five successive cases of chorea with repeated subcutaneous injections of magnesium sulphate. In every case a 25 per cent. sterile solution was used; the dose ranging from 0.01 gram (1/6 grain) per kilogram of body weight, i.e., 0.04 c.c. of the 25 per cent. solution at the beginning of the treatment, with a daily increase to 0.2 gram (3 grains) per kilogram of body weight, i.e., 0.8 c.c. of the 25 per cent. solution at the last injection. The actual amounts of the solution used daily were from 3 to 30 c.c.; they were given three times daily for from 10 to 15 days with the ordinary Record syringe in the back, loins and buttocks. In only one of the five cases was there a noticeable improvement after the series of injections, and in this one case the movements subsided gradually. Not only did the remaining four cases show no improvement, but they all had severe inflammatory reactions after the injections, and in one of them a marked albuminuria set in.

In marked contrast with Heiman's experience was that of Schroeder,²⁹ who used a 20 per cent. solution of magnesium sulphate subcutaneously in nine children, and two adults with severe chorea, with such good results that he recommended this method to be used in all hospitals.

SERUM TREATMENT OF CHOREA.—Goodman,⁴⁰ of New York, investigated the etiology of chorea and noted pathologically the involvement of the central nervous system; this suggested to him that the disease was infectious in nature and that the older methods of treatment had failed because the remedies had not reached the seat of the disease, so he began to use the patient's own serum intraspinaly. His technic is as follows:

- (1) Exclude syphilis and tuberculosis.
- (2) Rest in bed for several days without treatment in order to eliminate all previously used drugs from the system.
- (3) Blood, to the amount of 40–50 c.c., is drawn from a vein and centrifuged; this amount of blood will furnish about 15–20 c.c. of serum. Keep on ice till ready for use.
- (4) Lumbar puncture is performed and 15–20 c.c. of spinal fluid is withdrawn.
- (5) The serum is warmed to body temperature and from 10 to 18 c.c. are injected into the spinal canal; this is done slowly, the average time consumed being from ten to fifteen minutes.

(6) The patient remains flat on his back for an hour or two.

Of 30 cases treated by Goodman, according to his original report, 14 received one injection, 8 two, 5 three, and 3 received four injections. Of those receiving 1 injection, 12 were cured and 2 markedly improved; of those receiving 2 injections, 5 were cured and 3 markedly improved; of those receiving 3 injections, 2 were cured, 1 markedly improved, 1 slightly improved and 1 unimproved.

Porter,⁴¹ of San Francisco, reported before the American Pediatric Society his experience in the treatment of chorea by means of **intrathecal injections of horse serum**. Seven cases were injected intrathecally in the ordinary way, and one by the method which Mehrtens of the Stanford University employs in the treatment of cerebrospinal syphilis. (Mehrtens injects arsphenamin from six to twelve hours after an initial dose of horse serum of 0.5 c.c. is given in order to find out whether the patient is hypersensitive to horse serum.) Five of the patients received a second injection on the fourth, fifth and sixth days following the initial dose; these injections were followed by strikingly rapid improvement in most cases, but no case ceased to twitch absolutely within a week; in fact, only in the mild cases was the twitching absent in two weeks. Porter admits that, on the whole, his results do not encourage the hope that this is any advance on other methods of treatment, and he would only advise the treatment as of value in controlling very severe cases but not in moderately severe cases. Oscar Schloss, of New York, employed the same method in 12 cases but he could find no difference in the results from twelve control patients who did not get the serum. La Fetra is not enthusiastic over either this (horse serum) or the Goodman method.

Timme⁴² reports a case of severe infectious chorea in which nothing was done for the patient except **absolute rest**; after four days **lumbar puncture** was done and 25 c.c. of fluid under high pressure, removed. At the end of 24 hours the patient although still suffering from the tonsillar infection became normal as far as the chorea was concerned. There was no return of the chorea. Another child with rheumatism, tonsillitis and chorea was subjected to the same treatment, and in 24 hours after the withdrawal of the fluid the chorea partially

subsided, although the child still had rheumatism and tonsillitis. Thirty c.c. more fluid was withdrawn and the choreic movements ceased absolutely and permanently. Strauss of New York, in attempting to follow the same procedure as a therapeutic measure was unable to obtain uniform results. The fluid was not under high pressure. Morse and Floyd¹⁵ found no noticeable effect at any time on the choreic symptoms after lumbar puncture. Indiscriminate lumbar puncture in patients with chorea is not advisable; we have seen several cases in which the chorea became more violent after the lumbar puncture, the patients becoming more fidgety than they had been before.

Various **injections with vaccines** (stock, autogenous, typhoid) and **sera** are being used by different clinicians. The writer has obtained favorable results with typhoid vaccine intravenously, but inasmuch as his patients were also subjected to hydrotherapy and to the use of sedatives, he hesitates to attribute the beneficial results solely to the use of the vaccine. Good results are also claimed from the intravenous use of **gentian-violet** (J. W. Visser⁴³).

MECHANOTHERAPY—Kinesitherapy.—In the milder and in the more chronic forms of chorea, **mechanical treatment** by means of **suggestion** and **passive movements** at first, and later by means of **exercise**, has been worked out and systematized by Guthrie. The movements employed are very simple; at first slow and steady, like in Fränkel's method of treating tabes. Grossman⁴⁴ reports successful results following treatment by breathing, relaxation and reëducational exercises. He also begins with the simplest possible movements, and as the patient begins to show signs of improvement he orders more elaborate exercises.

HYDROTHERAPY.—Electrotherapy.—**Mild hydrotherapy** in the form of warm packs followed by cold spongings or warm tub baths lasting for one or two hours are sometimes very beneficial. Some physicians employ **electricity**, in the form of galvanism to the back, head and pressure points (if any can be found). We can see no rationale in this form of treatment and therefore do not recommend it.

TREATMENT DURING CONVALESCENCE.—In convalescence, gymnastic exercises, a change of scenery and a sojourn at the seaside or in the mountains will hasten to establish a cure.

INSTRUCTIONS TO PARENTS.—It is wise to impress upon parents the importance of **clothing** their children **properly**; not to consider lightly frequent attacks of *sore throat*; to pay greater attention to so-called *growing pains*; to remember that *night terrors*, *nervousness*, *clumsiness*, *restlessness*, and *incontinence of urine* may be the first symptoms of an impending attack of St. Vitus's dance, and lastly, that *rheumatism* and *chorea* both have a great tendency to recur.

Prognosis.—The ordinary outcome is a complete cure, but in rare cases a permanent loss of muscle power or a weakness of the mental faculties may remain. When these occur they are probably due to organic changes in the nerve centers, which have been brought about by the chorea. The mortality in Pineles's experience is 2-4 per cent.; in the cases of the British Chorea Committee the mortality was approximately 2 per cent.

The prognosis is much more favorable in children than in adults. It is more serious if the chorea follows immediately, or soon after any of the infectious diseases. In the very severe cases of chorea gravis (chorea

acutissima) the prognosis is very bad; most of the patients die within a week or two. A guarded prognosis must be given in cases complicated with severe mental symptoms, or definite organic cardiac lesions. The same is true of cases in which hyperpyrexia is a prominent symptom.

The prognosis is somewhat better in a recurrence than in a first attack.

When death occurs, it is usually due to cardiac failure or to exhaustion from insufficient nourishment and lack of sleep, or to injuries and infections sustained during the violent and incessant jactitations.

Pathology and Pathogenesis.—The most constant lesions found are endocarditis, the mitral valve being the one most commonly involved, fatty myocardium with Aschoff bodies (submiliary nodules in the wall of the left ventricle), and ulcerative endocarditis with metastatic emboli in other organs.

The changes found in the nervous system are as varied and as numerous as those who describe them.

Kopczynski could find no changes in the brain. Starr considers the disease a functional one and regards the pathologic findings in the nervous system as secondary changes. Gowers and others also believe these changes to be secondary in nature. Schiötz,⁴⁵ from a study of 211 cases, concludes the condition to be a neurosis which develops in those predisposed to it between the ages of eight and twelve, and that the inciting cause may be either toxic, infectious or emotional stress. Runge⁴⁶ points out that inasmuch as most of the psychoses occurring in chorea are similar to those seen in the exhaustion and infectious psychoses, certain weight must be given to the theory of the infectious origin of the disease. Bechterew also thinks that the disease is infectious, but whatever the infectious agent may be, it does not produce gross lesions in the nervous system.

Choreiform movements have been produced experimentally in animals by Rosenthal and by Angel Money⁴⁷ by injecting fluid into the carotids, as a result of which, capillary emboli were found in both the brain and cord. Stimulation of the motor cortex with chemical poisons, especially creatinin, also was followed by choreiform movements.

Dickinson⁴⁸ found as the most common pathologic lesion a hyperemia of the brain and cord with hemorrhages in that part of the brain which is supplied by the middle cerebral artery. Steiner, Hutchinson, Clarke and others are of the opinion that lesions in the spinal cord are responsible for the choreic movements; they base their opinion on the anemia and proliferation of connective tissue which they found in the upper part of the cord.

Changes in the axis cylinders of the peripheral nerves have been found by Elischer,⁴⁹ and changes in the muscles in "paralytic chorea" by Rindfleisch.

Horatio C. Wood believes, as a result of his experiments in animals, that the pathology of chorea is a change in the nutrition of the ganglionic structures of the entire cerebrospinal axis, and that this change in nutrition may in some cases fail to develop structural changes sufficiently marked to be detected by the microscope, while in other cases it may go on to the production of pronounced structural lesions.

Both Meynert⁵⁰ and Elischer⁴⁹ found hyalin degeneration in the nerve cells of the central ganglia. Flechsig describes similar changes limited to the anterior part of the lenticular muscles. Elischer, Jako-

wenko and others found concentrically situated, strongly refractive granules, the so-called "chorea corpuscles," in the vessel walls of the lenticular nucleus, but Wallenberg and his followers do not consider these characteristic of chorea, because these same "corpuscles" have also been demonstrated in diseases of the brain other than chorea.

Alzheimer⁶¹ reports changes in the corpus striatum and subthalamic region in two cases of chorea associated with sepsis; and in two cases of "rheumatic chorea" there were small foci of proliferated glia cells with rod-shaped cells, particularly near the vessels, and in the septic cases, heaps of cocci occluded the vessels. In the "rheumatic" cases there were similar foci of cells but no microorganisms could be demonstrated. He thinks therefore that septic and rheumatic chorea both depend on embolic foci, which are most frequently situated in the corpus striatum and subthalamic region.

Damaye⁶² believes chorea to be an organic brain disease due to an encephalitis. Comby⁶³ thinks that the disease is a neurosis of the same character as epilepsy, and that both of these are most commonly residua of an acute encephalitis. Chorea, he believes, is an expression of a mild, acute, curable infectious encephalitis, and that almost any infectious disease may provoke it. Gareiso⁶⁴ reports signs of encephalitis in 49 out of 50 cases of Sydenham's chorea; the organic signs, especially the Babinski toe phenomenon, varied from day to day, and as the chorea subsided, these evidences of "transient" encephalitis also disappeared. In the writer's experience the presence of a true Babinski toe phenomenon with the other usual evidences of pyramidal tract involvement is extremely rare in ordinary Sydenham's chorea.

The conception that chorea is due to an encephalitis is not new. As far back as 1864 Hughlings Jackson⁶⁵ spoke of chorea associated with fatal cases of acute rheumatic fever as "meningo-encephalitis," and localized the lesion "around the corpus striatum." Jackson's conception was confirmed two years later by the necropsy studies of Bastian, who placed the "principal lesions in the basal ganglia—chiefly the corpus striatum." The work of Jackson and Bastian was later confirmed by Poynton and Paine.

Various theories have been evolved to explain the mechanism of the choreiform movements themselves. Thus, Gowers, Bonnhoeffer and Foerster think that the cerebellum, or the tract of the superior peduncle is the starting point of the disease. Oppenheim apparently leaves the question open when he states "There can be no doubt that chorea is a brain disease, but we cannot say with certainty whether the lesions are localized in the central ganglia, the cerebral cortex, the cerebellum or in all of these." Conos,⁶⁶ drawing his conclusions from the case of a 77 year old woman with epilepsy and chorea, attributes the choreiform movements to lesions which he found in the lenticular nuclei. A critical analysis of this case, however, casts a great deal of doubt whether this observer's patient had what we would today designate as Sydenham's chorea. The same may be said of Libert's⁶⁷ case which showed at necropsy two small tumors in the dura overlying the frontal region. Perhaps the most ingenious theory is that of J. R. Hunt,⁶⁸ who believes that lesions involving the small ganglion cells of the caudate nucleus and putamen (neostriatum) give rise to chorea, because the function of this system of cells is inhibitory and coördinating, and loss of function in

these cells by disease gives rise to the wild and irregular movements.

As a result of careful clinical and pathologic research in diseases of the extrapyramidal system, Jakob⁶⁹ is led to believe that the motor disturbances of toxic, infectious (Sydenham's) chorea and of symptomatic chorea are due to focal changes in the striatum. He attributes the toxic infectious chorea which develops with especial frequency after articular rheumatism, partly to embolic, and partly to infiltrative and degenerative focal processes similar in character to those observed in the cases of chorea, which complicate epidemic encephalitis. Chorea gravidarum, he also believes to be due to embolic lesions in the blood-vessels of the striatum.

Summary of Etiology, Pathology and Pathogenesis.—The evidence presented in the previous pages, as to the etiology and pathology of chorea, shows that up to the present time there is no uniformity of opinion as to the pathogenesis of the disease.

The apparently contradictory etiologic and pathologic findings are probably due to the fact that the different writers, in describing chorea, confuse *Sydenham's* or *infectious* chorea with the *degenerative choreas*, and with the *symptomatic choreas*, as well as with the chorea-like movements observed in many cases of *psychogenic tic*.

Considering Sydenham's chorea in the sense that it is described in this chapter, the writer is justified in making the following statements:

(1) That there is more than abundant clinical evidence that rheumatism, chorea, and some forms of endocarditis are closely related to one another.

(2) That the gradual or sudden onset of the disease, its slow progressive course with occasional fever, its tendency to recurrences and to cardiac and arthritic complications, as well as the similarity of the psychic symptoms, when such exist, to those of the infectious or toxic psychoses, point to a probable toxic or infectious origin.

(3) That whatever this agent may be, whether infectious or toxic or both, it has hitherto not been demonstrated, but bacteriologic investigations seem to indicate that it is most probably a bacterium whose cultural characteristics are still to be determined.

(4) That the toxin or toxins elaborated by this bacterium has a predilection, in those predisposed, for nerve, joint and cardiac tissue, just like the toxic agent in what is known as Wilson's disease (see p. 533), has a predilection for the lenticular nucleus and the hepatic cells.

(5) That, while there is no doubt that chorea is a brain disease (encephalitis), the exact localization for the motor phenomena has not as yet been definitely determined, but that it is most probably due to involvement of the basal ganglia, especially the striatum and its connection with the extrapyramidal pathways.

Historical Summary.—The names of St. Vitus's dance, the dance of St. John, chorea minor, chorea major, and chorea germanorum have been used with varied meanings in regard to their significance. It appears that the Phrygian bacchantes, in their wild worship, were affected with violent automatic movements, accompanied by more or less disturbance of consciousness, and it is certain that the sect of the Suffi, in Persia, shortly after the origin of Mohammedanism, were accustomed in their sacred ceremonies to pass into a condition of wild excitement with danc-

ing, muscular spasms and general convulsions. About the year 1000, sect of the Suffi found numerous followers and imitators throughout Asia Minor, in Persia, Egypt and Greece. In Christian countries the so-called dance of St. John was already, at the time of the Crusades, an observed custom. It was not until the outbreak, in 1418, of a fresh epidemic of religious excitement in Strasburg that the term "dance of St. Veit" began to be freely applied to these religious disorders, because during this outbreak the chief magistrate of Strasburg ordered those afflicted with dancing mania to repair to the chapel of St. Vitus in Zabern, a village in Alsace, near Strasburg. The name St. Vitus appears to have had its origin from St. Veit, a boy who, born in Sicily, suffered martyrdom in the year 303 during the persecution of Diocletian, and whose body was carried from place to place, until finally it was buried in the cloister of Korvey.

Paracelsus called these epidemics "*chorea sancti viti*" and "*chorea lasciva*." The name, St. Vitus dance, is the only point of affinity between the old religious dancing manias and the "idiopathic" chorea of the present day. "Idiopathic" chorea was placed on a firm, scientific footing by the classical description of Sydenham. Many German writers speak of the affection in childhood as *chorea minor*, while the term *chorea major* or *chorea germanorum* is used to express affections resembling those of the hysterical epidemics of the Middle Ages. Sometimes the term "*chorea major*" or "*chorea germanorum*" is used to designate what we ordinarily understand to-day by "hysteria major."

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HUNTINGTON'S CHOREA

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Synonyms.—Hereditary chorea, Chronic progressive chorea, Choreic dementia, Huntington's disease, Degenerative chorea.

Definition.—A chronic progressive hereditary disease, appearing rarely before the end of the third decade of life, and characterized by irregular choreic movements, speech defects and gradual dementia.

Etiology.—SEX.—The prevailing opinion seems to be that both sexes are about equally affected, although Huntington thinks that the disease is more prevalent in males. Wollenberg's statistics bear out Huntington's claims.

AGE.—Cases of Huntington's chorea have been reported in childhood. Stevens reported a case which began in infancy, but it is questionable whether it was a case of true Huntington's chorea. Jolly saw a case in which chorea and epilepsy developed at the age of 9 years. Osler saw a case at the Johns Hopkins Hospital in an 18 year old individual. Hoffman, Peretti and others have reported cases which began in the second decennial period of life. Mackey had 2 or 3 cases below 30. These and similar cases are exceptions; the disease is preëminently one of adult life beginning most frequently in the middle period of life, or early part of the second half, between 30 and 40. It rarely commences in old age. It seems that the age of onset bears some relation to the course of the disease; there are certain strains of families in which the age of onset is earlier than in others. According to Heilbronner, when

the disease descends through several generations, it has a tendency to develop later and later in life.

HEREDITARY INFLUENCES.—The disease is transmitted from one generation to another; one generation may be passed over, or its members may suffer from insanity, epilepsy or hysteria instead of chorea. From the statistics at hand, it would appear that if children of choreic ancestors get through life without any manifestations of the disease, the thread is broken and the grandchildren and great-grandchildren may be assured that they will be free from the disease.

Muncie, a field worker for the Eugenics Record Office, was set to collect statistics on the disease (originally started by Jelliffe). She was able to construct 4 great pedigree charts containing 441 female and 521 male choreics—962 cases in all, and in addition 10 cases of Sydenham's chorea. The entire number of individuals studied was 4,370. The relatives, even those who did not have chronic chorea, had other nervous diseases. Thus, epilepsy occurred 39 times; infantile convulsions, 19 times; meningitis and encephalitis, 51 times; hydrocephalus, 41 times; feeble-mindedness, 72 times; Sydenham's chorea, 11 times; and ties, 9 times, mostly in one small family. These 962 cases originated from 6 or 7 ancestors who settled in eastern Long Island, south-central and south-western Connecticut, and eastern Massachusetts; from these localities the disease spread along the lines of immigration as far as the Pacific coast—one case having been found as far west as Los Angeles. The disease has been handed down in these families without a break through four generations. Among these families people of high mental accomplishment were by no means rare; they included legislators, professors, ministers, authors, one judge and one eminent surgeon. Some, however, broke down later in life, many of them showing lack of responsibility, immorality and a tendency to alcoholism.

Lewis¹ reports a case of Huntington's chorea in a man of 50, living in Allegany County, in the State of New York, the members of whose family have always recognized that they were liable to diseases of the nervous system. An older brother of the patient had a similar condition which was never diagnosed; he died insane. One younger brother has a similar condition, but not so marked; one sister is neurotic, but has neither motor nor mental symptoms. The mother died at 60 of pneumonia, after having suffered from similar symptoms, and it is believed, although not positively known, that the patient's maternal grandfather had the same trouble. One collateral female cousin has the disease in a very severe form; other cousins could not be traced.

De Castro reports two typical cases of the disease in two individuals with no history of heredity in either family, and they both have perfectly normal children. The writer has now under observation 2 cases, in neither of which any hereditary relationship, as far as chorea is concerned, can be ascertained although both give a bad family history of neuropathy.

The disease bears no etiological relationship to Sydenham's chorea.

Symptomatology.—**MODE OF ONSET.**—Huntington's chorea has, as a rule, a gradual onset. *Premonitory symptoms* in the nature of mild muscular twitchings, clumsiness in movements requiring coördination, and abnormal mental states may occur for years before the typical, irregular, coarse, jerky movements make their appearance. Emotional disturbances, traumata and pregnancy seem to be common exciting

causes. Horstman² reports a case in a man who developed the disease suddenly after he fell off a horse and sustained an injury to his head, but two years before the injury, without having shown any choreiform movements, his mental condition was very poor. He committed suicide, and on postmortem, typical lesions of Huntington's chorea were found.

OBJECTIVE SYMPTOMS.—The most striking part of the clinical picture, and the one without which the disease cannot be recognized, are the peculiar, sudden, tonic movements of the head, trunk and extremities. The movements are those of extension, flexion and grasping; there is a worm-like spreading of the fingers, somewhat similar to athetoid movements, but much more rapid in rhythm. The patients "pucker up" their lips, wrinkle their foreheads, dilate the nostrils, open and close their eyes, and roll their tongues. This continuous "play" of involuntary movements gives rise to most grotesque gestures, grimacing and peculiar speech. As a rule, the movements begin in one of the lower extremities and extend to the upper extremities and face, but they may remain confined to one extremity or to the face for a considerable period. Mental excitement and physical strain intensify the movements; during rest they become less marked, and usually, though not always, cease during sleep.

The patient can for a short period inhibit the movements by voluntary effort; he thus is able to eat, write, grasp an object, thread a needle and perform other movements requiring considerable coördination. The author had under his care a patient who, although subject to most violent movements, had no difficulty in shaving himself with an ordinary razor. This voluntary inhibition of the choreic limbs is often at the expense of increased activity in the muscles of the other limbs, and the volitional movements are executed, not regularly and continuously, but only at certain times, the choreic movements reappearing in the intervals.

In uncomplicated cases, muscle power is, as a rule, undisturbed, but the myotatic irritability is increased. In some but not in all cases, the continuous movements may be followed by considerable fatigue.

As the disease progresses the limbs become somewhat rigid, and the patients develop a peculiar gait during which the upper part of the body seems to advance ahead of the pelvis and legs with the trunk assuming a rocking movement, and the arms swinging from side to side. The legs seem to skip steps, and locomotion in general is carried out quickly and irregularly. The gait, often resembles that of a drunken man—"cerebellar gait." At about this time, a slight tremulousness of the hands is superadded to the choreiform movements; Romberg's sign may often be elicited. The face becomes expressionless and slight ptosis of the lids may be noticed. The writing, earlier in the disease, is tremulous and irregular but later it cannot be carried out at all.

The deep and tendon reflexes are usually normal at the beginning; as the disease advances they become somewhat exaggerated, and with the onset of the rigidity they become unusually lively, and cannot be elicited at all as the rigidity becomes very marked. The superficial reflexes are unaffected and there are no pathologic reflexes. The pupils react to light and in accommodation. The fundi are normal. The blood and cerebrospinal fluid show no pathologic changes. There are no objective sensory disturbances. The sphincters remain intact.

MENTAL SYMPTOMS.—There is a progressive weakening of the processes of perception, association, memory, attention and judgment. The patients are excitable, aggressive, and dull emotionally. Absent-mindedness is a characteristic and early symptom which is often wrongly interpreted as dementia. Delusions of persecution and phobias are more common than delusions of grandeur. The patients are rarely euphoric; they are more commonly depressed and show a marked tendency to commit suicide. Some of them are very loquacious and may have transitory attacks of mania; they are all very restless and suffer frequently from insomnia. As the disease advances they become disoriented as to time, place and person; all mental faculties are in abeyance, and the restlessness is replaced by apathy. The mental symptoms may precede or appear simultaneously with the hyperkinetic phenomena, but, as a rule, they are most marked during the terminal stages of the disease. There are some rare cases in which the mental state remains unimpaired.

Diagnosis.—The characteristic features of the disease are: (1) direct heredity; (2) hyperkinesia characterized by sudden, purposeless, jerky movements of the head, face, trunk and extremities; (3) onset in middle or later life; (4) gradual progression of the symptoms with no remissions, and (5) progressive mental deterioration. These five diagnostic features are usually found together, but in a good many cases some of them may be absent. Several members of an affected family, or two or more generations may show an atypical symptom-complex which will be specific for that family. The hyperkinesia rarely skips a generation, and is therefore considered a pathognomonic feature. In some families a psychic disorder of a hyperkinetic type, i.e., mania, may be a predominating feature.

There are thus various biotypes of the disease: there is a biotype without tremors but with mental weakness; another with tremors and no mental weakness; another in which the onset of the motor symptoms is early in life; another in which there is a considerable interval of time between the development of the mental symptoms and the choreiform movements. Different strains of families have different symptom-complexes, so that the age of onset, the degree of motor restlessness and the extent of mental degeneration will show family differences, and will enable one to recognize various biotypes of the disease.

DIFFERENTIAL DIAGNOSIS.—In general, it may be said that if the hereditary nature of the disease cannot be definitely established, the symptoms alone are not sufficient to distinguish the disease from simple chorea, unless one has an opportunity to observe the chronic progressive course later in the disease. The evidence of heredity is also diagnostic in the differentiation of this form of chorea from senile chorea due to arteriosclerotic and degenerative processes of the cortex, although the latter rarely begin before the age of 55.

Oppenheim and Remak report a form of "familial chorea" in two boys, sons of a woman suffering from a chronic form of hemichorea. Both boys became affected at the age of 8 with a typical chronic, progressive chorea which involved at first the lower extremities and gradually spread to the entire body, with a predominance of symptoms in the legs.

Some authors regard Unverricht's myoclonia similar to hereditary chorea, but this, in the opinion of Oppenheim, is not justifiable.

The differentiation between Huntington's chorea and paresis, when

he latter is associated with choreic movements, i.e., "choreatic paresis," may be very difficult. On the mere positive biological findings as to paresis, the diagnosis of Huntington's chorea cannot be excluded because the two conditions may coexist (Lowrey and Smith³).

Association with Other Diseases.—Huntington's chorea may be associated with epilepsy which may begin simultaneously with the chorea, or it may precede it by months or years. In one of Remak's cases epilepsy was present from the twenty-third to the thirty-first year and the chorea developed at 40. Diefendorf saw a case in which epilepsy developed at 17 and chorea at 53. In one of our cases at the Montefiore Hospital the chorea began simultaneously with the epileptic convulsions. Benedek and Goldenberg⁴ report a family in which Huntington's chorea was associated with migraine.

Treatment.—**PROPHYLAXIS.**—It has been suggested by Muncie and others that, owing to the hereditary influences, the State concern itself with the investigation of the progeny of every case of Huntington's chorea, and order sterilization of all those who already show symptoms, and to secure legislation that such of their offspring as show premonitory evidences of the disease shall not be allowed to reproduce.

TREATMENT OF SYMPTOMS.—The treatment is purely symptomatic. The **bromids, hyoscin, luminal, allonal, paraldehyde** and other sedatives in conjunction with hydrotherapy are employed for the alleviation of the choreiform movements, but the results do not seem to be very encouraging. The writer obtained no results from the use of arsenical preparations. It is important to take precautions to prevent these patients from committing suicide. With the onset of the muscular rigidity, the general helplessness of these patients presents very difficult problems in feeding and nursing.

Course, Duration and Prognosis.—The course of the disease is a very slow but progressive one; cases have been recorded whose duration was from one to two years (Gowers), but so short a duration is very rare. The disease is incurable and lasts several decades; it has very little tendency to shorten life. There are cases reported with recoveries, but it is doubtful whether they were cases of genuine Huntington's chorea. In so-called "choreatic paresis" if antiluetic treatment can be resorted to early and intensively, the prognosis should be comparatively favorable. Towards the end of the disease the patients become bedridden and succumb to some intercurrent disease or cachexia from lack of nourishment, bed sores and coma. Very many patients become despondent and commit suicide.

Pathology and Pathogenesis.—Lannois and Paviot⁵ describe a proliferation of the neuroglia and infiltration of the cortex with glia cells as the chief pathologic findings. Kattwinkel found atrophy of the supratangential fibrous layer and of the radiating fibers in the central convolutions with round cell accumulation around the Betz cells. Similar changes were also found by Rossi, Buck, and Spiller. Besta attaches great significance to the vascular changes. Stier⁶ and Müller⁷ think the disease due to congenital malformations of the motor cortex, on the basis of which the subsequent changes develop. The former also found diffuse degeneration of the spinal cord. Oppenheim, Hoppe, Greppin, Kalischer, Kronthal, and Facklam found disseminated military encephalitic foci in the motor cortex. Binswanger as well as McCarthy speak of

changes not unlike those of general paresis. Raecke⁹ found the greatest evidence of disease in the cortex of the central convolutions but there were no vascular changes. Collins⁹ found in one of his cases chronic parenchymatous degeneration of the motor cortex with slight degeneration of the pyramidal tracts of the spinal cord. Damaye¹⁰ found in two cases meningo-encephalitis with an intense neuronophagia in all stages. Kéraval-Raviot found in one case a meningo-encephalitis with round cell infiltration surrounding the smallest vessels.

These contradictory pathologic findings were not reconcilable with the clinical picture of a disease which seemed to run as uniform a course as Huntington's chorea does. The pathology of the disease, therefore, was not at all clear until about 1908 when with the more recent and better histologic methods available, Jelgersma succeeded in establishing a sound and scientific pathologic basis for the condition. Jelgersma's investigations were in the main soon confirmed by such competent observers as Alzheimer,¹¹ Pierre Marie and Lhermitte,¹² Margulis,¹³ Kleist, Pfeiffer,¹⁴ Kiesselbach,¹⁵ Hunt,¹⁶ the Vogts, Stern, F. H. Levy,¹⁷ Bielschowsky, and Jakob.¹⁸

A critical survey of the researches of these investigators would seem to justify the following conclusions: The pathologic basis of chronic progressive chorea is a very diffuse parenchymatous process involving the gray matter of the brain, particularly the corpus striatum (caudate nucleus), giving rise at first to a degeneration and later to a disappearance of the small ganglion cells with a relative freedom from involvement of the large ganglion cells. In the cases with mental symptoms



ILLUSTRATION 2.—HUNTINGTON'S CHOREA. Frontal Section. Myelin Fiber Stain. Atrophy of the Caudate and Putamen: Normal Pallidum. (Courtesy of Prof. A. Jakob.)

the cerebral cortex is also found to participate in the degenerative process; in most of these cases the pathologic process is most intense in the frontal lobe and precentral gyrus with the formation of a peculiar pseudonuclear gliosis above the Betz cell zone. Occasionally the parietal, temporal and occipital lobes may also be affected. In the areas involved there is a marked degeneration of the internal nuclear layer and of the lowest cortical layers (see illustrations 2, 3, 4 and 5, pp. 506-508).

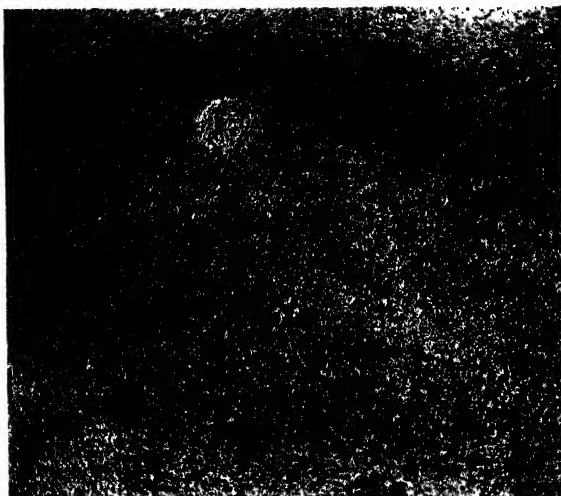


ILLUSTRATION 3.—STRIATUM. Nissl Stain. Microphotograph showing Degeneration and Disappearance of the Small Ganglion Cells and a Relative Increase of the Large Ganglion Cells. (Courtesy of Prof. A. Jakob.)

It is important to bear in mind that clinically as well as pathologically there is no difference between the hereditary and the non-hereditary cases of chronic progressive chorea.

As far as the pathologic process itself is concerned, there is histologically nothing specific about it. It is a purely degenerative process involving the parenchyma similar to that observed in chronic alcoholism and in the senile involutional processes such as senile dementia. The only feature of the pathologic process of chronic progressive chorea that may be said to be specific for the disease is the characteristic localization of the lesions.

Historical Summary.—The disease was first described in 1872 by George Huntington of Pomeroy, Ohio, at the time a practitioner on Long Island. It seems that the disease, as described by Huntington, has long been familiar to physicians practicing in districts adjacent to Long Island. Duglison¹⁹ quotes a letter written to him by Dr. Walters of Franklin, New York, giving a description of the malady almost identical with that of Huntington; it was then known to the laity as "magrums." Lyons, in 1863, published an article on chronic



ILLUSTRATION 4.—CHRONIC PROGRESSIVE CHOREA. Ten Years' Duration. Frontal Section. Myelin Fiber Stain. Advanced Atrophy of the Caudate and Putamen with a Mild Dysmyelination of the Globus Pallidus. (Courtesy of Dr. A. Jakob.)



ILLUSTRATION 5.—STRIATUM Nissl Stain. Microphotograph showing Complete Disappearance of the Small Ganglion Cells and Marked Degeneration of the Large Ganglion Cells. (Courtesy of Dr. A. Jakob.)

hereditary chorea in the *American Medical Times*, in which he described the disease as we see it to-day. Gorman, in his inaugural dissertation delivered before the faculty of the Jefferson Medical College, stated that the disease as described by Lyons was prevalent in certain parts of Pennsylvania.

Speaking of his personal experiences with this form of chorea (Huntington's chorea), Huntington,²⁰ in an address delivered before the New York Neurological Society in 1910, said: "Over 50 years ago, in riding with my father on his professional rounds, I saw my first case of 'that disorder,' which was the way in which the natives always referred to the dreaded disease. I recall it as vividly as though it had occurred but yesterday. It made a most enduring impression on my boyish mind, an impression every detail of which I recall to-day, an impression which was the very first impulse to my choosing chorea as my virgin contribution to medical lore. Driving with my father through a wooded road leading from East Hampton, L. I., to Amagansett, L. I., we suddenly came upon two women, mother and daughter, both tall, thin, almost cadaverous, both bowing, twisting and grimacing. I stared in wonderment, almost in fear. What could it mean? My father paused to speak to them and we passed on. Then my Gamaliel-like instruction began; my medical education had its inception. From this point on my interest in the disease has never wholly ceased."

Since Huntington's description of the malady, many instances have been recorded practically all over Europe and in various parts of this country. No cases seem to have been reported from Turkey, South America and the West Indies. While negroes seem to be comparatively free from Sydenham's chorea, Huntington's chorea has been met with in this race, associated with other nervous diseases, especially with epilepsy.

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OTHER FORMS OF CHOREA

Chronic intermittent chorea, p. 510—Chronic perennial chorea, p. 510—Senile chorea, p. 510—Post-hemiplegic chorea, p. 510—Hemiballismus, p. 510—Prehemiplegic chorea, p. 510—Chorea in encephalitis lethargica, p. 510—Hysterical chorea (*Chorea major*, *Chorea magna*), p. 511—Chorea natatoria—Chorea malleatoria, p. 511—Localized, isolated or partial chorea, p. 511—Tarantism, *tigretier*, p. 511—Dubini's chorea, p. 511—Electric chorea, p. 512—Bergeron's chorea, p. 512—References, p. 512.

Chronic Intermittent Chorea (Oppenheim).—In this variety relapses follow so quickly, and as the disease progresses, the intervals of freedom from symptoms are so short that the patients may be said to have chronic chorea.

Chronic Perennial Chorea (Oppenheim).—This form of chorea may last for years and even for a whole lifetime; it is very rarely seen in children. Oppenheim also recognizes a *chorea adultorum permanens*, which is distinct from hereditary or Huntington's chorea.

Senile Chorea.—This is a form of chorea appearing in old age and is usually due to vascular lesions in the striatum. It is a permanent condition and is very often, though not always, associated with mental symptoms. The latter are most likely due to the accompanying cerebral arteriosclerosis.

Posthemiplegic Chorea.—This rare form of chorea is met with in incomplete hemiplegias. The choreiform movements may be associated with a typical intention tremor, athetoid and associated movements in the uninvolved limbs; voluntary movement of the incompletely paralyzed limbs intensifies the choreiform movements. Posthemiplegic chorea is usually a hemichorea due to lesions in the posterior part of the thalamus, red nucleus, striatum and superior cerebellar peduncle. Extensive capsular lesions followed by secondary softening of the above enumerated structures may also give rise to this form of chorea. When the lemniscus participates in the pathologic process, the choreiform movements are accompanied by severe pain—"central pain," a characteristic symptom in the thalamic syndrome. Mitchell designated these cases as "painful choreas." Owing to the frequent involvement of the thalamus, these patients also present evidences of emotional instability,—"thalamic hyperaffectivity."

Hemiballismus.—Kussmaul has described under the name of "hemiballismus" a rare form of hemichorea characterized by violent, throwing, turning or twisting movements involving an entire half of the body. In the cases reported, the hemiballismus appeared several days after the onset of the hemiparesis. According to Jakob this disorder of motility is due to acute vascular lesions (hemorrhagic) in the *corpus Luysii*; he, therefore, designates hemiballismus as "*the syndrome of the corpus Luysii*."

Prehemiplegic Chorea.—This is a rare form of hemichorea observed in cases of slow bleeding into the thalamus. The choreiform movements precede the hemiplegia, and are best interpreted as an irritative phenomenon.

Chorea in Lethargic Encephalitis.—The choreic movements observed in this disease may occur very early in its course, or weeks and even

months after the acute symptoms of encephalitis have subsided. They may be generalized, unilateral, or segmental in distribution; they may vary in amplitude, and are occasionally rhythmical. A certain degree of hypertonicity is sometimes associated with the movements, giving rise to so-called "choreo-athetosis." According to Duncan they occur as a sequela to encephalitis in about one-sixth of the cases. They may be associated with parkinsonism and all its symptoms, or with typical myoclonic movements. When they occur during the acute stages of the disease the prognosis is very grave.

Hysterical Chorea (Chorea Major; Chorea Magna).—Hysterical individuals are frequently subject to general spasmodic movements, which at times simulate those of chorea. These movements are sudden, shocklike, rhythmical contractions, mostly noticeable in the hands and fingers, and not as severe as those in true chorea. Sometimes the involvement is more extensive and the movements are more violent. The movements are psychogenic in origin.

Chorea Natatoria—Chorea Malleatoria.—Chorea natatoria, in which the patients make movements with their hands as in swimming, and chorea malleatoria in which the movements are similar to those of beating a hammer on an anvil, are manifestations of hysteria, and have nothing to do with chorea.

Localized Chorea—Isolated or Partial Chorea.—There are rare cases of *localized, isolated or partial* chorea, in which the choreiform movements are limited to the lips, tongue, pharyngeal or laryngeal muscles. The movements have only a superficial resemblance to genuine chorea. They are probably psychogenic in origin. They are discussed in this chapter only because they are constantly reported in the literature as choreas, when, as a matter of fact, they are really forms of tic or habit spasm.

Tarantism—Tigretier.—Tarantism and tigretier are usually described as forms of chorea although they are both manifestations of hysteria. Tarantism is not known to-day. It is supposed to be due to the bite of the tarantula, and when the "disease" develops, periods of depression and stupidity make their appearance, after which, when the sound of a musical instrument is heard, the patients leap into the air and indulge in the wildest form of dancing and shouting which is continued until they drop from exhaustion.

Tigretier is a disease of modern times closely resembling tarantism; in fact, both of these conditions are in many respects like those forms of religious excitement which are accompanied by suspension of inhibitory control and disordered muscular movement. The "convulsionnaires" in France and the "jumpers" in Maine are good examples of individuals suffering from this type of motor disorder.

Dubini's Chorea.—Dubini described a form of chorea observed in northern Italy, which may appear at any age. It begins with pains in the head, neck or back; this is followed by electric-like, rapid, short muscle spasms, involving first one arm and one side of the face, later the homolateral leg, and lastly the other side. These shocks may be accompanied by unilateral epileptiform seizures. As the disease advances paralysis comes on in the affected extremities which soon becomes general. The muscles become atrophied and their electrical irritability is diminished. This form of chorea is very painful; there is

a marked hyperesthesia of the skin, the slightest touch of it producing the most violent contractions. The mind remains clear. High temperatures are common. After days, weeks or months, death ensues in coma or from cardiac failure. Very few cases recover. The disease is probably infectious in origin. On autopsy, meningo-encephalitis has been found.

Electric Chorea.—Henoch describes a form of chorea in which the muscular contractions differ from those in infectious chorea, in that they follow one another with lightning-like rapidity. The muscles of the shoulder and neck are most commonly involved. There is no change in the nutrition of the muscles and there are no sensory disturbances. The prognosis is good except that the contractions are so frequent and intense that the patients are incapacitated from work. Recovery takes place in a few days or weeks after the use of arsenic and hydrotherapy.

Bergeron's Chorea.—Bergeron described a similar disease except that it affected delicate and anemic children.

There is very little known of the etiology and pathology of these so-called choreas (Dubini's, electric, Bergeron's). The writer never saw a case belonging to this group but with the onset of the first epidemic of encephalitis lethargica, and before American neurologists became acquainted with the clinical picture of the latter disease, there occurred several cases which were at first thought to be forms of Dubini's or of electric chorea. These cases afterwards turned out to be myoclonic forms of epidemic, or lethargic encephalitis.

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PARALYSIS AGITANS

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Synonyms.—Paralysis agitans, Parkinson's disease, Shaking palsy.

Definition.—Paralysis agitans is an incurable disease appearing usually in the fifth decade of life, and characterized by the gradual onset of tremor, muscular rigidity and weakness, giving rise to a peculiar attitude, gait and facial expression.

Etiology.—FREQUENCY.—The disease is said to be more rare in Germany than in America. Berger¹ found 37 cases among 6,000 patients with nervous disease. Putzel² saw, during eleven years, 30 cases among 4,600 patients at the Clinic for Nervous Diseases in the Outpatient Department of Bellevue Hospital. König³ found in Siemmerling's Clinic

in Kiel, from 1901 to 1913, 23 cases among 5,000 patients with nervous disease. During the year 1914 there were in the wards of the Montefiore Home and Hospital, 30 cases of paralysis agitans among 161 patients with nervous disease. (Owing to the method of selecting patients for admission to this hospital, these figures are of no statistical value as to frequency.)

AGE.—The disease usually begins after forty years of age. In Gowers' 115 cases, two-fifths began between fifty and sixty, and about one-fifth in each of the two decades between forty and fifty and between sixty and seventy; it appears almost twice as frequently between sixty and seventy, on account of the lesser number of persons living during that decade. Occasionally it begins between thirty and forty, rarely under thirty or over sixty-five years of age. Gowers' series included 2 males beginning at seventy-three, 1 female at seventy-four, 1 case at twenty-nine (his youngest case). Hadden saw a case develop at twenty-five, Buzzard at twenty-one, Duchenne at nineteen, Berger at seventeen, Ballet-Rose at fifteen, Lannois at twelve, Weil and Rouvillois at ten. Ramsay Hunt⁴ reported 4 cases, 1 of which began at thirteen, 1 at fifteen, 1 at twenty-six and 1 at thirty. J. S. Bury⁵ reports 2 cases in a brother and sister at eighteen. Clerici-Medea reports 1 at twenty-eight and 1 at twelve.

SEX.—The disease seems to be more frequent in males than in females.

RACE.—Judging from statistical reports the disease seems to be more prevalent in the Anglo-Saxon than in the German race. It is very rare among negroes, and is said to be comparatively rare in Italy.

HEREDITARY INFLUENCES.—As a general rule, the influence of heredity in the development of the disease is not marked, although it may be a predisposing cause in many cases. Both Berger and Gowers found in 15 per cent. of their cases a history of the disease in more than one member of the family. Of Wollenberg's 19 patients 5 had a family history of mental and nervous disease. Oppenheim knew of one family in which two sisters developed paralysis agitans at an early age, a third becoming affected late in life with senile dementia with peculiar choreic movements of the tongue. In another one of his cases of paralysis agitans, a brother had the disease and a sister had bulbar palsy. He also saw one family in which three sisters had the disease. From his cases he is led to believe that the disease has a special tendency to appear in long-lived families. Clerici-Medea⁶ saw in one family 2 sisters with the disease, and in both it developed at an early age (*see* under Age). Bury (*see* under Age) is also of the opinion that certain families are more prone to it than others. Bonnhoeffer and Siehr saw 2 cases in one family. Lundborg⁷ reports 7 cases of paralysis agitans in three generations of the same lineage in one Swedish peasant family; besides these, 2 more cases existed in that family which were never diagnosed by a physician.

SOCIAL INFLUENCES.—Occupation, exposure to lead, brass and mercury, and one's station in life seem to have little or no influence on the development of paralysis agitans. Living in damp rooms, or prolonged exposure to wet and cold has been, in Putzel's cases, a most efficient etiological factor. Walz thinks the disease occurs only in individuals who have a deteriorated nervous system. J. M. Clarke suggests that the condition is one of premature senescence—abiotrophy of certain neuron systems, which may occur at any age.

In our cases at the Montefiore Hospital the patients affected with paralysis agitans were persons of good emotional equilibrium. Bad habits, dissipation, sexual excesses, overindulgence in food, alcohol or tobacco seem to have been no determining factor in our cases.

ANXIETY, WORRY, EMOTIONAL EXCITEMENT, FRIGHT, GRIEF, ANGER.—Prominent among the predisposing causes mentioned by most writers are anxiety, worry, emotional excitement, fright, grief and anger. Several cases due to fright developed among the inhabitants of Metz and Strasbourg during the Franco-Prussian war. Lorain reports the case of a seventeen-year-old girl frightened by the bursting of a shell in the cellar in which she had taken refuge during the siege of Paris; she immediately developed a tremor of the right arm which soon extended over the entire body, and at the end of five years she was still suffering from paralysis agitans. In the light of our experience in the late war, it would be interesting to know whether this was a case of genuine paralysis agitans or a "war neurosis."

TRAUMA.—Trauma with its accompanying psychic effects is well recognized as a predisposing cause. Walz's⁸ analysis showed, in 26 cases: general concussion in 6 cases; stabs and cuts in 7; burning and freezing in 1; sprains, fractures and twists in 4; and contusions in 8.

It was Charcot who first pointed out that physical injuries in general were frequently a predisposing cause in the production of the disease, and furthermore that it often began first in the part which was injured. Krafft-Ebing,⁹ however, found in his series of 110 cases, only 7 cases in which trauma could be attributed as a cause. He believes that fatigue due to overstrain in the muscles is the cause; he is led to this belief because in 50 out of 88 cases he found the disease had developed first in the right upper extremity, and this was the extremity utilized by the patients in the course of their daily tasks. This is the reason, he thinks, that unless trauma to the lower limb is the predisposing cause, the disease begins most commonly in the upper limbs.

Trauma may also determine the spread of the disease in a case which is already showing signs of it.

INFECTIOUS DISEASES.—Cases have been reported after typhoid fever, typhus, malaria, syphilis, epidemic encephalitis and the other infectious diseases, but it is doubtful whether these can be considered determining factors in cases of genuine or idiopathic paralysis agitans.

Symptomatology.—**CLINICAL HISTORY.**—*Mode of Onset.*—The usual onset of the disease is gradual. Prodromal symptoms, such as burning or rheumatic pains, weakness, numbness, girdle pains and local non-inflammatory swellings, frequently precede the characteristic symptoms of tremor and rigidity. These are sometimes erroneously considered manifestations of rheumatism. They are vague and wandering, their distribution more or less general or localized in the limb or limbs which later become affected with the disease. They may make their appearance years or months before the development of the other symptoms.

PHYSICAL FINDINGS.—*Tremor.*—Tremor is the first symptom of the disease in two-thirds of the cases; there are rare cases in which it is absent throughout the entire course of the disease—the so-called *paralysis agitans sine agitatione vel tremore*.

The tremor consists of slow, rhythmical oscillations, 4–7 per second, noticed chiefly in the distal ends of the upper extremities, the hands and

fingers. The movements take the form of flexion and extension, abduction and adduction of the fingers; flexion and extension, pronation and supination of the hands. They are usually within narrow limits, the thumb and index finger merely rubbing together as in "rolling pills" or "counting coins." The movements in the hand and forearm are less limited, and may sometimes amount to true shaking. The tremor is slight at the beginning and increases with the progress of the disease; it lessens in frequency as it increases in amplitude, the fine tremor of the early stage being quicker than the coarser tremor of the later period.

The most characteristic feature of the tremor as pointed out by Parkinson is that *it continues during rest*. It is present when the patient is lying down or standing, whether his hands are supported or hanging by his side. The character of the tremor is remarkably *stereotyped*; the same movements of flexion and extension occur in the same tempo and with the same amplitude of oscillation all the time. It may from time to time increase or diminish in its intensity, but only for a second or two after which it resumes its original character.

Voluntary active movements stop the tremor for a time; it ceases momentarily when the patient extends his hand or alternately opens and closes it. Momentary cessation of the tremor may be observed when he grasps an object or changes the position of his limbs, or when he is made to fix an object with his eyes. This temporary cessation is due to the general inhibiting effect of active movement.

During forced active movements or during slow but continuous movement, such as writing, the tremor persists or is increased; active movement associated with excitement also increases it. A patient with shaking palsy may be more comfortable when walking than when sitting or lying down, and may be able to walk long distances without being annoyed by the tremor. The tremor ceases in the limbs involved in an attack of hemiplegia, to return with the recovery from the hemiplegia. Variations occurring in the rhythm or rapidity of the tremor later in the disease are due to the effect of the accompanying muscular rigidity. The relation between tremor and rigidity is aptly expressed by Hughlings Jackson when he says, "Tremor is rigidity spread thin, and rigidity is tremor run together."

Passive movements may stop the tremor for a time or even altogether (Oppenheim), but it is then to some extent transferred to other muscles and is increased in the extremity which is not being manipulated. Concentrating the patient's attention, or a mere attempt to touch the shaking limb may also temporarily inhibit the tremor. The tremor always ceases during sleep and under narcosis.

The shaking may be confined to one arm or hand, or it may involve both arms, or an arm and leg on the same side, or all four extremities. In the hemiplegic form the facial muscles may also participate in the involvement. Pronounced tremor is generally less common in the lower than in the upper extremities. When the legs are involved the tremor is most noticeable in the calf muscles, but it may be seen in the thighs and even in the toes. Occasionally the muscles of the trunk and back may participate in the tremor, but it is almost never seen in the abdominal muscles. The head is not always spared; tremor of the lower jaw, of the muscles of the chin, lips and tongue is not uncommon. Gowers saw the tremor once in the orbicularis palpebrarum, and Westphal in

the lower facial muscles. In one of Oppenheim's cases the tremor of the eyelids was so intense that he could not make an ophthalmoscopic examination of the patient's eyes.

Rarely do the muscles of respiration participate in the tremor. Müller, Graeffner, Cisler and others report tremor in the vocal cords, and Rosenberg saw it in the vocal cords and velum palati. (Cf. Speech, p. 517.)

Muscular Rigidity.—Another characteristic symptom is muscular rigidity; its intensity bears no relation to the intensity of the tremor. As a rule, it sets in later than the tremor, and the patients may be conscious of it long before it can be elicited by the examiner. The facial expression, attitude, gait, and the slowness in the execution of voluntary movements are all due to this rigidity.

The rigidity is "cadaveric" and differs from that of pyramidal hemiplegia or paraplegia in that it is permanent and is not increased by the movement which elicits it. It produces a resistance in the limbs which does not vary whether the limbs are moved slowly or rapidly. Passive movement elicits a slightly "interrupted" rigidity of the muscles—the so-called "cog-wheel" phenomenon (Moyer)—not unlike the sensation experienced by one on "pulling a ratchet." Another peculiarity is that, in spite of its severity, the rigidity may be overcome on repeated passive movement, except in the older cases in which marked contractures have already resulted.

Attitude and Facial Expression.—As the disease advances it produces almost absolute fixation of the entire body causing the patient's head to be inclined forward, as if fixed to the trunk. The facial muscles having lost their emotional play almost entirely, the face looks as though it were covered with a mask. The back is curved in kyphosis—rarely in lordosis or scoliosis. The patient holds himself "rod-like," with arms abducted, the elbows flexed and the wrists extended, with the fingers either flexed at all joints or at one joint; at times—especially when the interossei are involved—the fingers may be extended, or the hand may assume a position similar to that in which a pen is held (Charcot's writing hand), or the fingers are dug into the palm of the hand (Déjerine's fakir hand). The thighs are flexed and adducted, the knees bent, and the feet held in a position of talipes equinovarus with a claw-like deformity of the toes.

Muscular Weakness.—Muscular weakness and rigidity usually come on together and are as characteristic of the disease as the tremor. The loss of muscle power varies much in degree: at first it is slight; it gradually increases, being usually greatest in the part in which the tremor developed first and most. This weakness must not be confused with the general weakness seen occasionally as a prodromal symptom. It never amounts to a total paralysis; it may precede the tremor or may begin simultaneously with it. Impairment and retardation of active movements in the disease are due to the muscular weakness and the rigidity, although they may be noticed even before the rigidity is very marked. Active movements are slow; the muscles do not seem to respond immediately to the will. The simplest movements are carried out sluggishly, as if with great deliberation; the more complicated the movement, the greater the sluggishness.

Gait.—In a typical case of paralysis agitans, when the patient is

asked to walk across the room, he begins hesitatingly, with short, shuffling steps, and hurries more and more until he reaches his destination—"festinating gait"; if at the point of destination he has no support or no object to grasp, he does not stop but keeps on running forward—"propulsion"; the patient is then said to "run after his center of gravity." There is sometimes a tendency to run backward—"retropulsion." Retropulsion may also occur on bending too far back, as in trying to take an object from a shelf. A similar phenomenon may be elicited on attempting to walk in a lateral direction, "lateropulsion." These peculiarities in gait can best be brought out by giving the patient a push in the desired direction or pulling his coat sideways or backward. Some patients walk much better backward than forward. Propulsion and retropulsion are much more common than lateropulsion. Oppenheim explains these disturbances in gait as due to the fact that the patient has great difficulty in bringing the groups of muscles which have been at rest or in a condition of tonic spasm rapidly into one of contraction or relaxation; he is unable to arrest the movement and is compelled to continue it in the direction once begun.

Loss of Normal Associated Movements.—When a patient with paralysis agitans is asked to swing his arms voluntarily, he has no difficulty in doing so no matter how much rigidity there may be in his muscles, but on walking he holds his arms stiffly side by side, and the natural swinging of the arms, as seen in a normal individual, is absent. An attempt at this "natural swing" of the arm can even be noticed in a patient with hemiplegia due to a lesion of the pyramidal tract; careful examination of such a patient will show that he cannot swing his arm voluntarily when asked to do so, but when walking, no matter how much spasticity or paralysis there may be in the arm, the tendency to swing it will be at once noticeable. The diminution or absence of normal associated movements in paralysis agitans can also be readily demonstrated when the patient is asked to "make a fist"; the normal extensor "kick" of the wrist is lost; the quick abduction of the thumb and the spreading of the fingers on opening the hand quickly, seen in a normal individual, are also absent in a patient with paralysis agitans. The loss or diminution of normal associated movements of the arms in walking, and on closing and opening the hand is regarded by Hunt as an important sign of what he designates as "pallidal palsy." This sign is helpful in differentiating paralysis agitans from the "functional" as well as from "organic" types of paralyzes due to pyramidal tract involvement.

Speech.—The voice in paralysis agitans is weak and whining; speech does not begin until some time has elapsed after the thought has been created. There is no modulation in the voice; after the patient has once begun to speak, the words roll over each other. These changes in speech depend upon the amount of tremor and rigidity in the muscles of the larynx and of articulation.

The study of the larynx in paralysis agitans is difficult on account of the presence of false tremors which may be very misleading. As has been pointed out, a genuine parkinsonian tremor may be present in the vocal cords as well as in the other muscles of the body. Graeffner¹⁰ has shown that at times this tremor may be on the same side as the tremor of the upper extremity, or it may be on the opposite side. In 80 cases he found 21 in which there was a tremor of the entire larynx of the same

tempo as the tremor of the body; in 27 the tempo was different; in 32 there was no tremor of the true vocal cords. In 8 cases without tremor of the body, tremor of the larynx was observed in 5 cases. In all cases presenting a laryngeal tremor the latter was most evident in the open position of the vocal cords; in 12 of the cases the adductors were involved. Cislér¹¹ found in 75 per cent. of his cases a "cadaveric rigidity" of the cords.

Eyes.—There is, as a rule, no true paralysis of the ocular muscles and no nystagmus; there are no ophthalmoscopic changes in the nerve heads or in the retina. Oppenheim noted in one case paralysis of convergence which he attributes to a tonic contraction of both abducens muscles. Slowing of the extraocular movements is common, and oculomotor paralysis has been described (Saint-Leger, Debove and Neumann). In two of Oppenheim's cases of unilateral paralysis agitans there were oculopupillary symptoms on the affected side; in another unilateral case he also noticed a positive von Graefe. Moczutowsky (cited by Oppenheim) lays stress on the fact that when the eyes are opened, the frontal muscles may sometimes be in a condition of tonic contraction so that the folds on the forehead disappear gradually instead of suddenly. We repeatedly observed this phenomenon, especially in advanced cases. Involvement of the extrinsic as well as of the intrinsic ocular muscles with irregularity and inequality of the pupils was very frequently observed in the parkinsonian syndromes encountered during the recent epidemics of "lethargic" encephalitis. Loss of reaction to accommodation with retention of the light reflex was unusually common, and sluggish reaction to light and even complete loss of the latter with retention of accommodation (Argyll-Robertson pupil) was not infrequent. The writer has also seen several cases with complete loss of reaction to both light and accommodation. The finding of these ocular changes in cases of parkinsonian syndrome in which syphilis could be excluded was a helpful aid in differentiating "encephalitic" or "post-encephalitic" paralysis agitans from genuine or idiopathic paralysis agitans, in which, by the weight of authority, no such changes occur.

Bulbar Symptoms.—Dysarthria, dysphagia, salivation and drooling are frequent, especially in advanced cases. Bernhardt noted in one case forced laughter similar to that seen in pseudobulbar palsy. Oppenheim once saw excessive salivation associated with an uncontrollable discharge of mucus from the nose. Owing to the diffuseness of the pathologic process in cases of epidemic encephalitis, bulbar symptoms are unusually common in the parkinsonian syndromes associated with that condition. The presence of bulbar symptoms early in the course of a case of paralysis agitans would ordinarily speak in favor of an "encephalitic" or "post-encephalitic" parkinsonian syndrome.

Reflexes.—The superficial and deep reflexes are always present, rarely increased; later in the disease when the rigidity is marked they may be difficult to elicit. There is no Babinski or any of its modifications present. False clonus due to tremor may be observed. Oppenheim has been able to elicit true clonus in a few exceptional cases. Tileston¹² also observed true clonus without other signs of pyramidal tract involvement. Graeffner has found the pharyngeal reflex absent in 20 out of 84 cases and markedly diminished in 9 cases. Mendel's observations in reference to the loss or diminution of the Achilles reflex were not

borne out in 28 cases examined by Graeffner, although this reflex was found to be modified in a number of the cases; these modifications, he believes, were more in conformity with the general findings in the senile and arteriosclerotic.

Huet-Alquier and others regard exaggeration of the deep reflexes as the usual condition. Our cases, in the absence of complications, never showed any abnormality in the reflexes.

In unilateral paralysis agitans abnormal associated movements in the toes of the healthy foot, when the patient moves voluntarily the affected foot, are not uncommon.

Sensation.—Acute pain, except as a premonitory symptom is, as a rule, not found in uncomplicated cases of paralysis agitans, although the French school has described a painful form of the disease which they call “forme douloureuse” (L’Hirondel). Some patients complain of a feeling of stiffness, burning or cold throughout the disease. They seem to be very uncomfortable at night because, on account of their rigidity, they cannot turn around in bed to change their position, and in some cases the paresthesias may be so distressing that the pressure of the bedclothes cannot be tolerated. Except in the cases in which the thalamus participates in the pathologic process there are no objective sensory disturbances.

Muscles.—The muscles show no change in size; late in the disease, when the patients have reached the stage of general helplessness, atrophy due to disuse and contractures may be seen.

As a general rule the electrical irritability of the muscles to both currents remains normal. Borgherini found a delay in the latent period of muscular contraction and a lessened irritability of the muscles and nerves to the electric current. The weight of authority is against these changes.

Skin.—Hyperidrosis is common. The skin of the hands may be smooth, erythematous and shining; occasionally it is edematous, and is designated by the French as “main succulente.”

Joints.—The joints are usually not affected, although French writers have suggested that there might be true parkinsonian arthropathies. Oppenheim thinks that these joint affections are probably combinations of paralysis agitans with arthritis deformans.

Sphincters.—In typical cases there is no involvement of the sphincters. If the subject of the disease is well advanced in years there may be complications, such as a hypertrophied prostate in the male, or a prolapsus uteri in the female, which for mechanical reasons may produce rectal or bladder disturbances.

Gastro-intestinal Tract.—Owing to the sedentary habits and lack of exercise these patients have poor appetites and suffer from constipation.

Laboratory Findings.—*Blood and Blood Pressure.*—In uncomplicated cases there are no changes in the blood picture or in the blood pressure.

Urine.—Phosphaturia is common; the sulphates have been found decreased in amount. The urea nitrogen is normal.

Cerebrospinal Fluid.—The cerebrospinal fluid shows no pathologic changes.

SPECIAL FINDINGS.—*Psychic Symptoms.*—Parkinson in his original essay mentions no mental symptoms as constituting a part of the clinical picture. According to Ball (cited by Putzel), paralysis agitans and in-

sanity are associated more frequently than is commonly believed. König found many cases of hypochondriasis, melancholia and paranoid conditions among his patients; rarely, if ever, was euphoria present; senile dementia and terminal deliria were quite common.

In general it may be said that in most cases the intellect remains unimpaired. The distressing symptoms toward the end, with their general helplessness, make the patients peevish, irritable and depressed. In our experience, no matter how severe the disease was before the patients became bedridden, they all seemed to be good-natured and satisfied with their lot. When mental symptoms are present they are characterized by emotional depression, agitation and psychic pain. Many patients have hallucinations generally referable to the organic sensations and to the sense of touch; their delusions are somatic and self-accusatory or paranoid in nature; some of them have memory defects with little or no mental confusion (Jackson, Free and Pike¹⁸).

Diagnosis.—The slow, progressive development in the fifth decade of life of a chronic painless disease characterized by tremor, muscular rigidity, a mask-like face, statuesque and rod-like station, with a festinating gait, is diagnostic of a typical case of paralysis agitans. There are, however, many atypical cases which may need prolonged observation before their true nature can be determined.

DIFFERENTIAL DIAGNOSIS.—*Multiple Sclerosis.*—Paralysis agitans is distinguished from multiple sclerosis by the fact that in the latter the tremor is coarser, not as rhythmic, and occurs only during the performance of a voluntary movement, i.e., an intention or "action" tremor, which is associated with nystagmus, optic nerve changes, a scanning speech, exaggerated tendon reflexes, absent or diminished abdominal reflexes, and bladder disturbances. If the paralysis agitans begins with weakness and rigidity but without tremor, the typical attitude of the head, trunk and limbs with the diminution or loss of normal associated movements will be diagnostic, but when the disease is unilateral and not associated with tremor, a correct diagnosis may be impossible.

General Paresis (Dementia Paralytica).—In paresis the tremor is not rhythmic, and does not persist during rest. The absence of the characteristic parkinsonian attitude and gait, and the presence of mental changes with positive biologic findings in the blood and cerebrospinal fluid will make the diagnosis clear.

Arteriosclerosis of the Brain and Cord.—Involvement of the striatum in cases of cerebral arteriosclerosis may produce a clinical picture resembling paralysis agitans. The presence of the usual clinical signs of diffuse cerebral vascular disease with a history of repeated attacks of cerebral vascular accidents giving rise to pseudobulbar palsy, and exaggerated deep reflexes associated with a positive Babinski sign or its modifications, and a spinal type of sensory disturbances, when the cord is involved, will be diagnostic criteria. The cases of so-called "*senile tremor*" are most likely also due to vascular lesions in the striatum; it may be impossible to distinguish clinically these from cases of genuine paralysis agitans.

Cerebellar Disease.—Cerebellar disease may give rise to lateropulsion or propulsion with tremor, but the coarseness of the latter combined with ataxia, hypotonia and dyssynergia will serve to distinguish between cerebellar disease and paralysis agitans.

Chorea.—The jerky, coarse, irregular character of the movements of chorea can hardly be confused with the tremor of shaking palsy.

Wilson's Disease.—The juvenile form of paralysis agitans may sometimes be confused with Wilson's disease (progressive bilateral lenticular degeneration). The coarseness of the tremor, the early onset of dysarthria and dysphagia with the characteristic mental symptoms and the very rapid course in the acute cases with fever and emaciation, will speak in favor of Wilson's disease.

Hysteria.—The presence of hysterical sensory manifestations, the hysterical mental make-up, the age and sex of the patient, and the nature of the tremor, which is irregular, of great amplitude, with a tendency to appear in attacks, influenced by suggestion and hypnotism, will aid in differentiating hysteria from paralysis agitans.

Traumatic Neuroses.—These are sometimes followed by a tremor, attitude and gait similar to that observed in paralysis agitans, and inasmuch as the latter occasionally follows trauma, a perplexing diagnostic problem may arise. The mental attitude of a patient with a traumatic neurosis engaged in litigation; together with the criteria enumerated under the heading of hysteria with the lack of progression of the symptoms will be in favor of the diagnosis of a traumatic neurosis.

Arthritis Deformans.—Spiller reports a case of paralysis agitans diagnosed as arthritis deformans, until a parkinsonian tremor appeared in one of the limbs. The presence of arthritic changes with the rigid attitude assumed by these patients on account of the pains together with the inability to determine whether or not there exist any changes in the normal associated movements, may make the differentiation between paralysis agitans and chronic rheumatism with tremor almost impossible.

Tremors of Metallic Poisoning.—The tremors seen in chronic metallic poisoning will be distinguished from the tremor of paralysis agitans by the history of occupation or exposure, the mode of onset and the course of the disease. (For further differential diagnosis, see section on Tremors, p. 468.)

Complications.—Symptoms of focal disease of the brain and spinal cord due to arteriosclerosis may frequently be associated with paralysis agitans, but inasmuch as such focal lesions may in themselves produce a parkinsonian tremor with rigidity, it is at times impossible to ascertain whether the condition is merely symptomatic of paralysis agitans, or whether it is a complication.

Berger has seen sudden attacks of transient hemiplegic weakness, and Gowers has known of occasional sudden attacks of transient general powerlessness occurring in paralysis agitans without any lesions being found in the brain to explain them. These are probably due to temporary spasm of the cerebral arteries—"intermittent cerebral claudication."

Early mental failure with loss of memory may be a complication of the disease. Gowers has once met with convulsions resembling those of epilepsy as a complication in a woman 59 years old, in whom the convulsions and the paralysis agitans began at the same time. Buzzard has reported a case in which there was a semi-cataleptoid condition of the limbs. The combination with tabes dorsalis is very rare, as is that with Basedow's disease. Luzzato and Lundborg have seen paralysis

agitans associated with symptoms of *myxedema*. Osnato¹⁴ reports a case of paralysis agitans and *myopathy* occurring in an uncle and nephew, with evidences of internal glandular disturbances in the latter. J. Roux¹⁵ saw a man 71 years of age with paralysis agitans complicated with an *acquired myotonia*.

Clinical Forms.—There is a tendency in modern medicine to consider paralysis agitans as being not a distinct clinical entity, but a syndrome with a definite characteristic complex of symptoms which may be due to various pathological conditions. With this view in mind, J. Ramsay Hunt,⁴ of New York, recognizes three types of the disease: (1) the presenile and senile; (2) the symptomatic type; and (3) the rare juvenile form. Paralysis agitans without tremor or without rigidity and all the other various irregular and incomplete manifestations of the disease, the so-called "*formes frustes*," he considers subdivisions corresponding to variations in the symptomatology. He believes that all these are forms belonging to the paralysis agitans group, and that they are related to each other clinically because they all present the chief symptoms of the syndrome (rigidity and tremor), but that they present differences which are due to different localization and different kinds of lesions.

The juvenile type of paralysis agitans he considers a pure system disease, due to a slowly progressive atrophy of the motor neurons of the globus pallidus mechanism, while the presenile, senile and symptomatic forms are dependent upon senile and vascular changes in the course of the same mechanism.

JUVENILE TYPE.—The juvenile type is characterized by the unusual frequency of familial incidence, the early age of onset, the more rapid progression of the symptoms, the comparatively early involvement of the bulbar muscles, the rarity of the form without tremor, and the infrequency of subjective sensory symptoms.

HEMIPLEGIC TYPE.—The hemiplegic type is not very common. In many of the cases reported as belonging to this type there was merely a predominance of symptoms on one side, but the facial expression and attitude were characteristic of typical cases of the disease. Juarros¹⁶ reports a case in which the tremor and rigidity were exactly confined to one side of the body and there was no sensation of heat or perspiration on that side; the reflexes were normal. When the rigidity precedes the tremor in these unilateral cases, they may simulate a case of slowly developing hemiplegia; the same is true when the involvement in one limb alone persists for any length of time. Marshall Hall distinguished a *hemiplegic* and a *paraplegic* type, and Berger added a *monoplegic* form. Gowers thinks that these should not be considered types, that they are merely prolonged stages of the disease, which later have a tendency to become generalized.

PARALYSIS AGITANS SINE TREMORE.—This is not a very uncommon variety. The tremor may be very slight or entirely absent; in these cases the diagnosis depends upon the rigidity, facies and attitude.

TYPES WITH VARIATIONS IN THE TREMOR—FORMES FRUSTES.—In some cases the tremor may be marked and constant in one part of the body, and be brought out in another part only on movement. In the latter cases the tremor may simulate that of multiple sclerosis (*see Diagnosis*, p. 520).

There are also rare cases in which tremor is present and rigidity slight or absent; in these, unless the tremor is very marked and typical, the condition may be impossible to distinguish from essential tremor, hysteria, or any of the other neuroses.

PARALYSIS AGITANS WITH HYPOTONIA.—Förster¹⁷ described a form of paralysis agitans with hypotonia, i.e., tremor without rigidity, and Krämer¹⁸ saw a rare "paralytic" type of paralysis agitans—typical paralytic disturbances without tremor and without rigidity. Hunt thinks that Krämer's and Förster's cases are vascular in type and not a system disease, not unlike the flaccid hemiplegias.

TYPES WITH PECULIAR ATTITUDES.—This type is not very common; in some the head may be inclined backward, or to one side as in torticollis, or rarely the entire body may be in extension instead of flexion (Charcot).

FORME DOULOUREUSE, FORME RHEUMATISMALE.—In these cases pain of a dull character over the joints or limbs may be so marked as to be the predominating symptom in the clinical picture.

SYMPTOMATIC FORMS.—Paralysis agitans occasionally follows true hemiplegia, being limited to the limbs first paralyzed. It is questionable whether these cases can be considered true paralysis agitans or complications of hemiplegia in which the tremor resembles paralysis agitans. This form has been noticed in lesions of the cerebral peduncles and of the basal ganglions due to neoplasms, inflammation and arteriosclerosis.

A good example of symptomatic paralysis agitans was seen by the writer in a man of 43 who was admitted to the Montefiore Hospital, complaining of fainting spells, weakness and poor memory, all of which set in three years before admission and following the death of one of his daughters. His previous history was of no significance. The only positive findings on examination were: Slight parkinsonian tremor with rigidity of the left arm and leg, and a tendency to a mask-like face with slight evidences of hyperthyroidism. All laboratory findings, including ophthalmoscopic examination, were negative. He was diagnosed as a case of unilateral paralysis agitans, most likely of the idiopathic variety. Prior to his admission to the Montefiore Hospital, he had been going around the various hospitals and clinics in which his condition was diagnosed as hysteria. After three months' stay at Montefiore, while up and about, and without any other complaints than those on admission, he suddenly became deeply comatose. He remained in deep coma several hours when he developed a severe general convulsion during which he died. Necropsy showed a large glioma involving the entire right temporal lobe; the neoplasm had flattened out the lenticular and caudate nuclei and optic thalamus on that side without invading these structures. The right ventricle was also partially collapsed and the septum lucidum of the third ventricle was pushed to the left.

PARALYSIS AGITANS AND EPIDEMIC (LETHARGIC) ENCEPHALITIS.—The paralysis agitans type of epidemic encephalitis is characterized by a fairly acute onset of the classical symptoms of parkinsonism—within two or three days after the acute onset of the encephalitis. Generally speaking, the tremor is less constant and less marked; sometimes it is overshadowed by the massive muscular rigidity, giving rise to "paralysis agitans sine tremore"; when present it is more coarse than the typical tremor of genuine paralysis agitans; it tends to be localized to the

tongue, face, one or more extremities or to a segment of an extremity. In the early stages of the disease the tremor rarely involves all extremities. The same is true of the rigidity. The symptoms of parkinsonism may disappear as the acute symptoms of encephalitis subside, but more often there is a tendency to progression; this tendency is greater in the cases in which the parkinsonism appears late in the disease, or as relapses after apparent recovery. The period of apparent well-being between the subsidence of the acute symptoms and the onset of the parkinsonism may vary from a few days to one or more years. This interval is usually longest in cases of subacute and chronic encephalitis. The usual history obtained from these patients is, that they had an attack of epidemic encephalitis with gradual recovery, after which they developed symptoms of paralysis agitans and entered on a stage during which they may, or may not have become lethargic again. This form of the disease would seem to depend on a renewal of the inflammatory process or on an actual lighting up of the old lesions (Economo,¹⁹ Globus and Strauss,²⁰ Schaller and Oliver,²¹ McKinley,²² Souques²³).

An interesting and also diagnostic feature of "post-encephalitic" parkinsonism is that many of the patients show a diurnal variation in their motor activities, and develop sleep disturbances in the nature of a "reversal of sleep," as well as various disorders of breathing, such as grunting, hyperpnea, dyspnea, constant yawning and all sorts of respiratory tics. These phenomena are rarely, if ever, observed in uncomplicated "idiopathic" paralysis agitans.

Another diagnostic feature is the occurrence of choreiform, myoclonic and dystonic movements in addition to the characteristic parkinsonian tremor and rigidity; this, too, is never observed in genuine paralysis agitans.

In contrast to the cases of genuine paralysis agitans, patients with "post-encephalitic" parkinsonism have a great tendency to develop various psychic disorders; these are most common in the younger patients and are characterized by a complete change of personality. Disobedience, excessive irritability, unprovoked fits of temper, cruelty, destructiveness, and even kleptomania have been observed. An important feature of the mental disorder in these individuals is that the change in personality or disposition is seldom accompanied by any marked degree of intellectual impairment. Beverly and Sherman^{24, 25} report two cases in which the patients (one, a boy of 11, and another of 15) developed behavior disturbances of a "post-encephalitic" type, prior to the development of the parkinsonian syndrome. One of the boys developed the syndrome six months, and the other two years after the appearance of the mental symptoms. The author has now under observation a 40 year old man with "post-encephalitic" parkinsonism with mental reactions typical of dementia paralytica.

Most cases of "post-encephalitic" parkinsonism show, in contrast to idiopathic parkinsonism, pupillary changes (*vide* p. 518) and residual signs pointing to extensive cranial nerve involvement, the facial nerve being most frequently affected; in many of these cases the emotional innervation of the affected side of the face shows more impairment than the voluntary innervation.

Treatment.—GENERAL.—Paralysis agitans being an incurable disease, the treatment is necessarily *symptomatic*. The patient is to be main-

tained in good general condition; mental excitement and physical strain are to be avoided; a carefully selected diet, very light exercise and mild hydrotherapeutic measures are recommended. In the later stages of the disease the patient should be kept in as comfortable a position as possible, and all measures should be employed to prevent contractures. Charcot pointed out that these patients usually feel better driving in a carriage or in a railway train; this has led to the construction of "fateuils trepidants," chairs which, by their continuous oscillations, give ease and comfort.

ELECTROTHERAPY.—Some claim that a weak faradic current, frequently interrupted, diminishes the tremor. Oppenheim recommends the use of electric baths and gentle passive movements. Whatever benefits may be obtained from electrotherapy are probably due to the psychic effect on these patients.

GYMNASTIC EXERCISES.—Exercises to relax the rigid muscles allowing the various members of the body to fall against gravity are recommended by Friedländer.²⁶ During these exercises overstrain must be avoided. Swift²⁷ of Boston reports improvement after graduated slow exercises given to the arms, hands, legs and toes for fifteen minutes, three times a day.

HYDROTHERAPY.—**Warm baths** are useful to control the rigidity. Several patients at the Montefiore Hospital, in the later stages of the disease, who had been rendered entirely helpless by the rigidity, were kept for two to three hours in a continuous bath of body temperature. The results were striking; some of the patients who had to be fed for months before this treatment was instituted could feed themselves after staying in the bath for several hours; the improvement, however, was only temporary.

MEDICINAL TREATMENT.—**Bromids** alleviate the restlessness and the feeling of anxiety. **Veratrum viride, cannabis indica, codein, opium,** and the various **alkaloids** of the **belladonna** group have been employed to lessen the tremor. The drug most commonly used and with the best temporary results is **hyoscin hydrobromate**; it may be given in fresh preparations by mouth, or preferably subcutaneously; during prolonged administration its toxic effects must be borne in mind although it seems that patients with this disease can tolerate unusually large doses (gr. 1/100), 0.0006 gram, three times a day, and can take it for a considerable period without any ill effects. Buss reports good results in "post-encephalitic" parkinsonism with daily injections of 5–10 c.c. of a 1/2 per cent. solution of **acriflavin** for from six to eight days. Erb was the first one to recommend **arsenic**; various preparations of this drug, **arsenious acid** and **Fowler's solution** by mouth, and **salvarsan, neosalvarsan** and **arsphenamin** intravenously, and **cacodylate of soda** intramuscularly have been employed. The writer had occasion to use these different arsenical preparations in a large number of cases of idiopathic and "encephalitic" paralysis agitans, and with the exception of two cases which were proven to be definitely syphilitic, his results were disappointing. Hohman reports good results from the use of **scopolamin hydrobromate** in doses as high as 1/50 grain (0.0012 gram) four times a day. Kennedy, Davis and Hyslop²⁸ could influence favorably the muscular rigidity in "encephalitic" parkinsonism by the administration of the fluid extract of **gelsemium** in doses of 7 minims

(0.42 c.c.) three times a day, or **gelsemin hydrochlorate** 1/30 grain (0.002 gram), by mouth three times a day; these favorable effects persisted as long as the drug was used—in one case, over three months. In some of the cases cumulative effects followed the use of the drug; these consisted of heaviness of the eyelids, diplopia, languor and confusion, they disappeared within 24 hours after its withdrawal.

Thyroid Extract, first suggested by Lundborg, seems to have no effect. The same may be said of **parathyroid** and of **pituitary extract**. Gordon, of Philadelphia, claims that his patients were relieved by the administration of **calcium lactate**. **Non-specific protein therapy** (typhoid vaccine intravenously), **sodium nucleinate**, **milk injections** and **autoserum** (intraspinaly) are being employed extensively. Here and there one encounters a case in which the tremor or rigidity, or both, are temporarily improved following treatment by these methods, but how much of the improvement is due to these measures and how much to the rest in bed that these patients are subjected to during treatment is problematical.

The writer has had personal experience with practically all these methods, and he is convinced that no treatment has much influence in arresting the progress of the disease. Gentle massage and passive movements with the judicious use of hydrotherapy and the administration of hyoscin in large doses frequently lessens the tremor and rigidity and makes the patient feel better; this applies to the "encephalitic" as well as the idiopathic types of paralysis agitans. In view, however, of the utter hopelessness of the disease, one is fully justified in using any and every prescribed method of treatment, if for no other rational purpose than for the psychologic effect that the employment of any well regulated plan of treatment has on the mental state of an unfortunate individual afflicted with a disease as disabling and hopeless as that of paralysis agitans.

Course and Prognosis.—The course of the disease is progressive, slow and chronic; it may take from fifteen to twenty years before the patient becomes so bent that he must be confined to bed. The tremor may for many years be limited to one extremity, usually the arm. It may take from one to three years before the leg is involved on the same side as the arm; or the leg and arm may become affected simultaneously. More rapid extension of the disease is not very common. There is usually a gradation in the severity of the tremor and rigidity in the different parts of the body, proportionate to the duration of the disease; there are, however, so many variations in the order of extension that it is impossible to foretell the future of a given case. This is especially the case in "encephalitic" parkinsonism.

The prognosis as to life is good. The sudden onset of bulbar symptoms in the "encephalitic" cases is quite common; the writer has seen several of these in which the patients while apparently comfortable, suddenly developed bulbar symptoms from which they died; many of the subacute and chronic cases terminated in this manner. Remissions may occur in the early periods of the disease, in the "encephalitic" as well as in the idiopathic cases; these may occur without, as well as with treatment. Sometimes a remission is followed by an aggravation of the disease. The course and prognosis of the symptomatic cases will naturally depend on the primary disease at the basis of the paralysis agitans syndrome.

Owing to the fact that paralysis agitans affects individuals well advanced in years, cerebral vascular accidents followed by paralysis frequently occur in the course of it. Attacks of apoplexy followed by hemiplegia lead to a cessation of the shaking in the paralyzed limb, but it generally reappears. Collet, cited by Oppenheim, noted a case in which the tremor ceased suddenly on one side of the body, although no paralysis had appeared. The tremor has also been observed to cease before death.

French clinicians claim that the prognosis in the "rheumatic" form is comparatively good.

The disease may last from 10 to 30 or more years. Death is usually due to some intercurrent complication, cerebral hemorrhage, decubitus, pneumonia, or general cachexia.

Pathology and Pathogenesis.—Parkinson in his masterly essay "On the Shaking Palsy" gives it as his opinion, which, he emphasizes, is merely "conjecture, and not based on pathologic evidence," that "the proximate cause of the disease is in the superior part of the medulla spinalis; and by the absence of any injury to the senses and to the intellect, that the morbid state does not extend to the encephalon." Since then, the pathologic basis for the disease was sought in various parts of the body.

Inasmuch as the predominating clinical signs are referable to the muscles, these were thought to be the site of the disease. So that we find that as far back as 1862 Skoda described fatty degeneration of the muscle fibers in Parkinson's disease; other observers corroborated these findings, and for a time there was a tendency to regard the disease as a form of myopathy.

With the acquisition of our knowledge of the rôle played by the glands of internal secretion in the body chemistry, many investigators focused their attention to these glands, and very soon paralysis agitans was attributed to dysfunction of the various ductless glands. Normal parathyroids being apparently the regulators of neuromuscular activity, Lundborg advanced the hypothesis that paralysis agitans might be due to disease of these glands. Similar theories were advanced for the thyroid, adrenals and even the hypophysis. The pathologic proof offered by the adherents of these theories was not convincing, so that the consensus of opinion to-day is, that whatever changes in paralysis agitans the endocrine glands and muscles may show, they are not to be regarded as pathogenetic factors in the disease.

As the number of postmortem examinations of cases of paralysis agitans began to increase, and with the advent of the more recent methods of neurohistologic examination, it became more and more clear that the cause of the disease must be sought in the nervous system. Various components of the nervous system began to be regarded as the site of the morbid process. Thus we find in the literature reports of involvement of the brain, spinal cord, spinal ganglia and even of the peripheral nerves. The lesions found, however, were not constant, and in the interpretation of their significance the question arose as to whether they were primary, or secondary and due to senile and vascular changes. It was not until the last decade of the last century that our knowledge of the pathology of the disease began to be crystallized, and the morbid process definitely established as degenerative in nature, and located in the basal ganglions.

It would seem that there exist to-day two schools of thought as to the pathogenesis of paralysis agitans. The one, represented by the Vogts, Lhermitte^{29, 30}, Hunt⁴ and others, considers the causative lesion of idiopathic paralysis agitans to be in the globus pallidus. The other, led by Tretiakoff³¹ and supported by Pierre Marie, thinks that the disease is due to lesions in the substantia nigra.

According to A. Jakob³² idiopathic paralysis agitans is anatomically dependent on a fatty degeneration of the neural parenchyma (the large ganglion cells) (see illustrations 6 and 7). The process involves chiefly

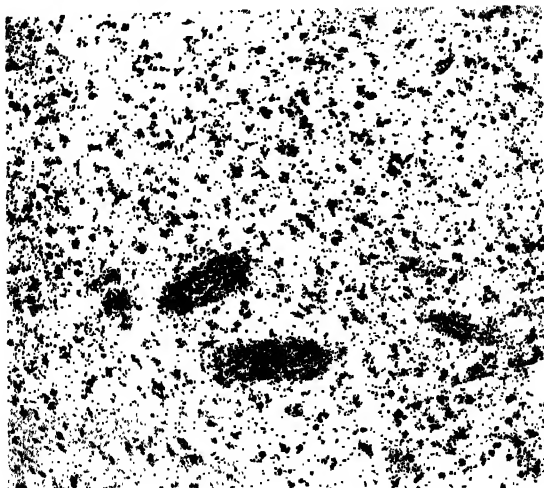
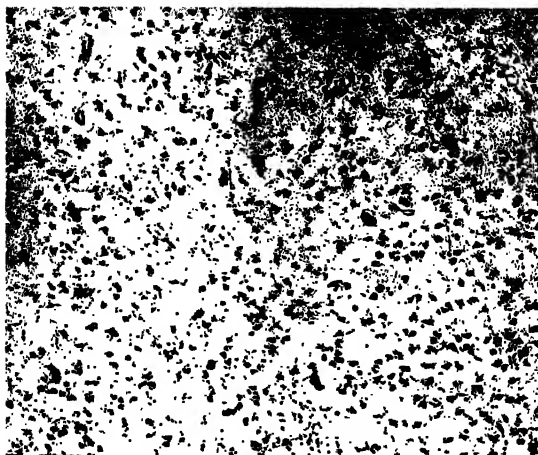


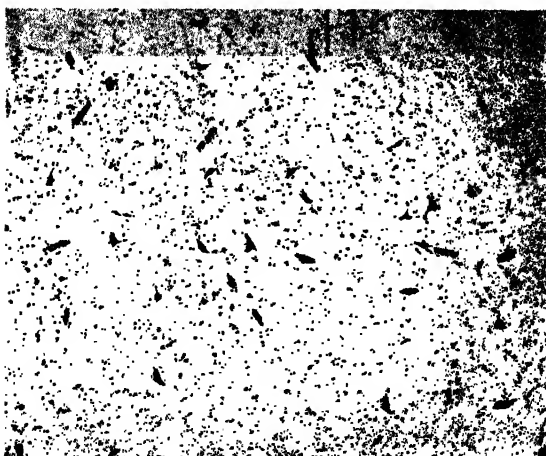
ILLUSTRATION 6.—PARALYSIS AGITANS. Fatty Degeneration of the Striatum. Herxheimer Stain. (Courtesy of Prof. A. Jakob.)

the striatum and in milder forms the globus pallidus (see illustration 8). The pathologic process resembles very closely that observed in senile involutional processes. The lesions of the parkinsonian syndromes (see illustrations 9 and 10) due to cerebral arteriosclerosis (see illustration 12) (Foerster's arteriosclerotic muscle rigidity) and syphilitic brain disease (see illustration 13, *A* and *B*) (vascular or dementia paralytica) are also situated in the striatum and pallidum and in some instances in the substantia nigra (see illustration 11, *A* and *B*). In these cases, the vascular changes can be differentiated only with difficulty from typical arteriosclerosis, but usually the more intense vascular infiltration and the proliferative changes in the cells of the vessel walls point to the syphilitic nature of the process (see illustration 13, *A* and *B*).

According to McAlpine³⁴ the pathologic basis of idiopathic paralysis agitans is a lesion of the globus pallidus or possibly of the subthalamic structures with which the former is connected, whereas "post-encephalitic" parkinsonism is usually due to a lesion (degenerative) in the substantia nigra. The constant affection of the zona compacta of the substantia nigra in "post-encephalitic parkinsonism" is emphasized by practically all



A



B

ILLUSTRATION 7.—A. NORMAL STRIATUM. NISSL STAIN. B. NORMAL GLOBUS PALLIDUS. NISSL STAIN. (Courtesy of Prof. A. Jakob.)



Pall.

ILLUSTRATION 8.—GENUINE PARALYSIS AGITANS. Myelin Fiber Stain. Frontal Section Showing Mild Dysmyelination of the Globus Pallidus and Ansa Lenticularis (*Pall. La*), and Mild Status Cribratus and Dysmyelination of the Putamen (*Put.*). (Courtesy of Prof. A. Jakob.)



ILLUSTRATION 9.—METENCEPHALITIC PARKINSONISM. Frontal Sections. Myelin Fiber Stain Showing Marked Degeneration of the Pallidum (*Pall.*) Ansa Lenticularis (*Al*) and Corpus Luysii (*Cl*). (Courtesy of Prof. A. Jakob.)

modern authors, although Jakob,³² who also claims that the substantia nigra involvement stands in the foreground, reports one case in which the striopallidary degeneration exceeded the manifest but receding changes in the substantia nigra. He also points out that the cortex is usually spared by the pathologic process.



ILLUSTRATION 10.—METENCEPHALITIC PARKINSONISM. Frontal Section. Myelin Fiber Stain Showing Marked Degeneration of the Substantia Nigra (*Sn*). Photograph. (Courtesy of Prof. A. Jakob.)

Hohman,³⁵ as a result of a careful clinico-anatomic study of eleven cases of parkinsonism associated with or following encephalitis, summarizes his conception of the entire process as a chronic progressive degenerative inflammation which is frequently associated with changes in the nature of a premature marasmus. The process is a diffuse one involving the entire central nervous system, although occasionally areas are found in which the lesions are so slight that they might easily be overlooked. In the chronic cases the large motor cells seem to show a relative immunity to the process; this, however, is not invariably so, because in some areas, notably in the mesencephalon (basal ganglia, optic thalamus), some of the large cells are severely affected. One of Hohman's cases showed, in addition to the involvement described above, changes in the cerebral cortex and in the dentate nucleus. From the point of view of intensity of the pathologic process the structures may be said to be involved in the following order: substantia nigra, striatum, pallidum and midbrain ganglia, dentate nucleus, cerebral cortex, medulla oblongata and spinal cord. The lesions were invariably symmetrically bilateral, even though there may have been some differences in the extent of the symmetry. Clinically, the cases showed that in some of them the parkinsonism began directly with the onset of the encephalitis and kept on progressing gradually but uninterrupted; in other cases there was a relatively free interval between the onset of the encephalitis and the development of the parkinsonism. On purely anatomico-pathologic

grounds the reason for the difference in the mode of onset of the parkinsonism in these two groups could not be explained satisfactorily. Hohman is, therefore, inclined to attribute this latency in the appearance of the parkinsonism to a regression of the inflammatory process. The entire pathologic process seems to be in favor of a gradually progressing chronic inflammation, because the step-like progression is not

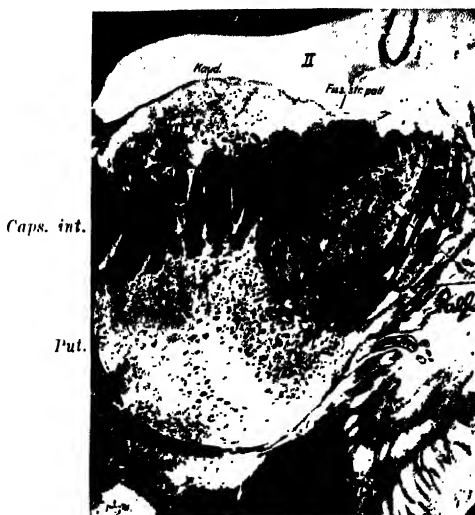
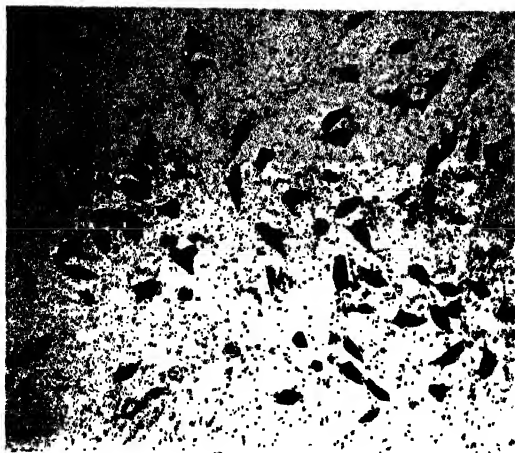


ILLUSTRATION 12.—ARTERIOSCLEROTIC MUSCLE RIGIDITY. Arteriosclerotic Foci in the Striatum (*Put.*) and Pallidum (*Pall.*). Myelin Fiber Stain. Photograph. (Courtesy of Prof. A. Jakob.)

evident in the clinical picture in most of the cases, even though here and there one does encounter a case with an apoplectiform type of progression. Hohman also emphasizes the fact that in view of the diffuseness of the pathologic process it is extremely difficult to correlate the clinical symptoms of "encephalitic" parkinsonism with any definite localization of the lesions.

In three cases of "post-encephalitic" paralysis agitans studied microscopically by McKinley and Gowan,³⁶ minimal changes of the globus pallidus but massive lesions of the substantia nigra were found. Scattered lesions found in other parts of the brains were so insignificant that the authors attached no importance to them. They believe that the syndrome of paralysis agitans may occur without a significant lesion in the globus pallidus, though they do not intend to convey the idea that this nucleus never plays a rôle in parkinsonism. The destruction of the substantia nigra is, in their opinion, the essential factor in the production of the syndrome. The discrepancies in the findings of the different authors as to which of the basal ganglia show a preponderance of involvement, is attributed by McKinley and Gowan³⁶ to the fact

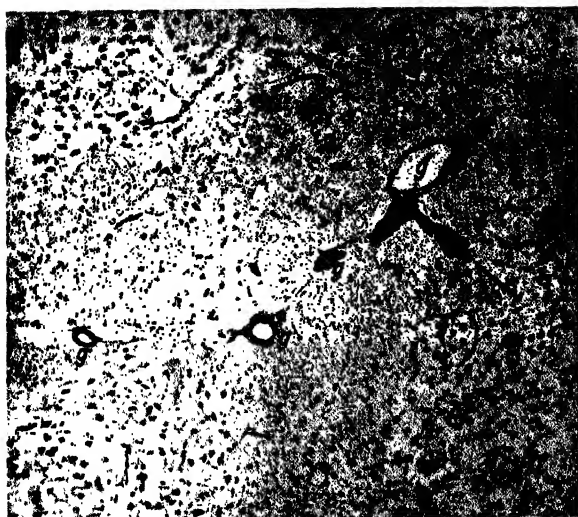


A



B

ILLUSTRATION 11.—*A*. NORMAL SUBSTANTIA NIGRA (ZONA COMPACTA). Nissl Stain. Microphotograph. *B*. Substantia Nigra in a Case of Metencephalic Parkinsonism; (*g*) Bloodvessel with a Mild Lymphocytic Infiltration. (Same Stain and Magnification as *B*.) (Courtesy of Prof. A. Jakob.)



B

ILLUSTRATION 13.—A. SYPHILITIC MUSCLE RIGIDITY. Large Focus (*H*) in the Putamen (*Put.*); Secondary Atrophy of the Pallidum (*Pall.*); *Kaud.*=Nucleus Caudatus; *l*=Nucleus Lenticularis; *Nr.*=Red Nucleus; *Caps.* B. Microphotograph of the Striatum of the Same Case Showing Syphilitic Changes in the Blood Vessel Walls (*g*) with Degeneration of the Surrounding Tissue. Nissl Stain. (Courtesy of Prof. A. Jakob.)

that up to the present no one had undertaken a detailed micrometric evaluation of the quantitative amount of cell destruction in these regions.

Conclusions.—From what has been said thus far, and from a critical survey of the literature, it would seem that there is as yet no unanimity of opinion as to the pathology and pathogenesis of the syndrome of paralysis agitans. Although it must be borne in mind that the final solution of this problem is yet to come, nevertheless the writer believes that there is abundant evidence to justify the following statements:

1. Uncomplicated idiopathic parkinsonism (genuine paralysis agitans) is due to a degenerative process (closely allied to that of "senescence") which is predominately localized in the striatum and pallidum, although lesions have also been found in the substantia nigra and in the corpus Luysii (A. Jakob²⁸).

2. Parkinsonism associated with senile dementia (senile muscle rigidity with dementia) is due to degeneration of the basal ganglions and of the cerebral cortex.

3. Arteriosclerotic muscle rigidity (parkinsonism due to cerebral arteriosclerosis) is due to arteriosclerotic vascular changes in the same localization as idiopathic paralysis agitans.

4. Syphilitic muscle rigidity is due to the same pathologic process as arteriosclerotic muscle rigidity, except that the vascular lesions are syphilitic in nature.

5. Parkinsonism in epidemic encephalitis is due to a diffuse degenerative parenchymatous process with occasional evidences of inflammation and infiltration, which, as the process becomes more chronic, also undergoes degeneration. The process involves predominately and regularly the zona compacta of the substantia nigra and, to a lesser extent, the pallidum, and least the striatum. In some cases it may also involve the cerebral cortex, pons, cerebellum, medulla and spinal cord.

Historical Summary.—The disease was first described by James Parkinson, a member of the Royal College of Surgeons, in 1817. The original description was in an octavo monograph entitled "An Essay on the Shaking Palsy" and was published in the same year by Sherwood, Nelly & Jones, London. Parkinson's definition of the disease is as follows: "Shaking Palsy (Paralysis Agitans). Involuntary tremulous motion, with lessened muscular power, in parts not in action and even when supported; with a propensity to bend the trunk forwards, and to pass from a walking to a running pace: the senses and intellects being uninjured." This definition practically covers the symptomatology of the disease as we know it to-day, more than one hundred years after the original essay was published. Parkinson's keen power of observation and clinical sense is also well illustrated by his masterly discussion of the differential diagnosis between paralysis agitans, chorea and other diseases characterized by the presence of disturbances of motility. Our clinical conception of the disease to-day remains practically the same as when Parkinson published his description of it; the only advance made was in its pathology. It was not until the last three decades during which increasing knowledge of neuropathology began to focus the attention of investigators to the basal ganglions as the possible site of the morbid process of the disease. It remained for the epidemics of encephalitis during the last seven years to further increase our knowledge of the pathophysiology of these centers, and their relation to the so-called

extrapyramidal systems, and thus establish a fairly reasonable pathologic basis for paralysis agitans and the motor disorders allied to it. It remains for the future to establish this basis more firmly, with the possibility of evolving a better therapy than we now have in our attempts to relieve the unfortunate sufferers from so disabling a disease as paralysis agitans.

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WILSON'S DISEASE

History and definition, p. 533—Symptomatology, p. 533—Diagnosis, p. 534—Clinical types, p. 535—Treatment, p. 536—Pathology, p. 536—References, p. 538.

Synonyms.—Bilateral progressive lenticular degeneration, hepatolenticular degeneration.

History and Definition.—In 1912 S. A. K. Wilson¹ described an extrapyramidal motor disorder, familial but not hereditary in nature, occurring usually in individuals toward the latter part of the second decade of life, pursuing an uninterrupted, chronic, progressive course, varying in duration from a few months to several years, and terminating fatally. Wilson's original paper was based on six carefully observed cases, although one case of this disease had been described by Frerichs of Berlin as far back as 1854, two cases by Gowers in 1888 under the title of "tetanoid chorea" and one by Anton in 1908 under the name of "dementia choreoasthenica."

The disease is characterized clinically by tremor, rigidity, dysarthria and progressive mental deterioration, and pathologically, by a progressive degeneration of the lenticular nuclei, and a peculiar type of hepatic cirrhosis.

Symptomatology.—1. TREMOR.—Tremor is present in almost every case, although it may vary in intensity in different cases. At the onset of the disease the tremor is fine, limited in range, and affects only the distal ends of the extremities; it is regular, rhythmic, from 4-8 oscillations per second, increased by physical or mental exertion, and can be voluntarily inhibited for a brief period. As the disease advances, the tremor becomes more extensive, and toward the end, involves the entire body. The tremor is parkinsonian in type, with occasional choreiform movements of the trunk and limbs during active voluntary efforts. In most of the cases reported, it appeared first in the right hand and was most noticeable when the patients attempted to write. Nearly as often it appears in the arms and legs simultaneously, less frequently in both arms alone, and in a few cases in the tongue. Although tremor is one of the characteristic signs of the disease, Borsari and Bianchi² have recently reported a case, proven by necropsy, which, though far advanced, showed no tremor.

2. RIGIDITY.—Another constant and characteristic feature of the disease is the rigidity or hypertonicity of the muscles. It is progressive in nature and involves all the muscles of the body except those of the eyes, although Stöcker³ in 1913 reported a case in which the external ocular muscles were involved. As the disease advances, the rigidity gives rise to contractures, which eventually lead to permanent deformities; these are most noticeable in the fingers, hands, toes and feet. The

face assumes a fixed and immobile expression. At times the contractures of the facial muscles of expression lead to an abduction of the angles of the mouth and a separation of the lips, giving the patient a silly, almost idiotic appearance. Although there is no true paralysis and no localized muscular atrophies, there is great difficulty in carrying out voluntary movements; this is due to the rigidity of the muscles and to general weakness. Owing to the rigidity of the muscles of the lower limbs, walking is extremely difficult; and late in the disease may become impossible. In many cases the rigidity and contractures are associated with tonic and clonic muscle spasms.

3. **DYSARTHRIA AND DYSPHAGIA.**—Difficulties in speech and swallowing are prominent features in the clinical picture. The dysarthria is characterized by a slurring of the consonants, and a "cutting off" of the last syllables. Toward the end of the disease there may be a total anarthria. The dysphagia usually appears simultaneously with the dysarthria. Deglutition is so impaired that even liquids are swallowed with great difficulty; the saliva accumulates in the mouth, and there is considerable drooling. It is important to bear in mind that although the patients may be unable to utter a sound there is neither paralysis of the soft palate nor of the tongue. The dysarthria and dysphagia are due to the rigidity of the muscles of speech and deglutition.

4. **MENTAL SYMPTOMS.**—These are as characteristic of the disease as the physical symptoms. They have been reported in almost every case; in some cases they were insignificant, in others very marked. Most of the patients showed a marked narrowing of the mental horizon, docility and childishness. In the chronic type of the disease there is euphoria, associated with restlessness, emotional instability and a tendency to explosive laughter. The terminal dementia, emphasized by all authors, is, according to Wilson, not a true dementia. These patients never show agnosia or apraxia or any of the defects of intelligence usually noted in senile dementia, general paresis and dementia precox. The intensity and progression of the mental symptoms bear no relation to the physical condition of the patient.

As in all pure extrapyramidal diseases there are no changes in the deep or superficial reflexes, and there are no pathologic reflexes. Some of the patients complain of generalized pains throughout the body and of painful cramps in the muscles of the limbs, trunk and neck. There are no objective sensory disturbances. The pupils and fundi are normal. There is no nystagmus, no cerebellar symptoms and no sphincteric disturbances.

Cirrhosis of the liver is a constant feature. In his original description of the disease Wilson pointed out that the cirrhosis was peculiar in that it gave no symptoms during life. Subsequent writers confirmed this observation. In view of the more recent methods of determining hepatic function, it will be interesting to note in the future whether cases of progressive lenticular degeneration will not show evidences of disturbed liver function. In this connection it is of interest to note that Homén's⁴ patients suffered occasionally from nausea; one of them also had frequent attacks of vomiting. In two of Wilson's cases there was a history of an attack of jaundice several years before the appearance of the symptoms referable to the nervous system.

Diagnosis.—The occurrence of a familial disease in the latter half of

the second decade of life, characterized by the presence of tremor, rigidity, contracture, difficulties in speech and swallowing with a progressive dementia, and the absence of symptoms referable to the pyramidal tracts, will, as a rule, offer no difficulties in reaching a correct diagnosis. The disease bears only a superficial resemblance to *multiple sclerosis*; the absence of nystagmus, disc changes, pyramidal tract signs with the preservation of the abdominal reflexes, as well as the rarity of remissions and the peculiar mental symptoms, will serve to distinguish Wilson's disease from multiple sclerosis. The presence in an elderly individual of bilateral pyramidal tract signs, paralysis of the palatal and lingual muscles, sphincteric disturbances, alternating attacks of spontaneous laughter and crying, together with a history of repeated attacks of cerebral vascular accidents, will distinguish *pseudobulbar palsy* from Wilson's disease. Greater difficulty may, however, be encountered in distinguishing between Wilson's disease and *juvenile paralysis agitans*; the early appearance of dysphagia and dysarthria, the presence of mental symptoms and the familial character of the disease will speak in favor of Wilson's disease as against juvenile paralysis agitans.

Clinical Types.—Wilson recognizes two clinical forms of the disease: acute or subacute, and chronic. The symptoms are practically the same in both forms. The acute or subacute form is marked by short duration (4–13 months), an irregular rise in temperature, and rapid emaciation; the entire clinical picture is that of a progressive toxic or toxi-infectious process. The chronic variety may last from 2½ to 7 years, the average duration being about 4 years. Except for the presence of fever, rapid emaciation and the shorter duration there is no difference in the clinical course of the two varieties. Relapses and remissions are extremely rare, but they may occur. The prognosis is invariably fatal.



ILLUSTRATION 14.—CUT SECTION, Showing the Entire Liver to be Cirrhotic; the Cirrhosis is Multilobular, not Bile-stained and Divided by Connective Tissue Hyperplasia. (Courtesy of Drs. F. J. Farnell and A. M. Harrington, and the *Journal of Laboratory and Clinical Medicine*.)

Treatment.—The treatment is entirely symptomatic. Sooner or later the mental symptoms become so marked that the patients must be committed to institutions for the insane.

Pathology.—On autopsy, the liver presents a most striking appearance—a typical “hobnail” liver (see illustration 14). The hepatic involvement is in all cases far advanced; the organ is strikingly small, nodular and firm, and never bile-stained. In all the “Wilson” livers, attempts at regeneration are shown by the active separation of the bile ducts in the connective tissue bands, and also by the mitotic division of the liver cells resembling a formation of irregular masses of cells, in which the architecture of the liver lobules is lost. The spleen is usually hypertrophied but shows no other evidences of disease. In some of Wilson’s cases the thyroid showed regressive and proliferative changes.

The most characteristic change, however, is the bilateral, symmetrical degeneration of the lenticular nucleus (see illustration 15), chiefly of



ILLUSTRATION 15.—BRAIN OF A CASE OF PROGRESSIVE BILATERAL LENTICULAR DEGENERATION. Horizontal Section Showing Almost Complete Destruction of the Left Lenticular Nucleus and Partial Destruction of the Right Lenticular Nucleus; the Caudate Nucleus is Much Smaller on the Left Side. (Courtesy of Drs. F. J. Farnell and A. M. Harrington, and the *Journal of Laboratory and Clinical Medicine*.)

the putamen and less of the globus pallidus. The caudate nucleus may be atrophied. The external capsule may be involved, but the internal capsule is intact, and the thalamus, save for the loss of the striothalamic fibers and a thinning of the lamina medullaris externa, is normal.

The degree of degeneration in the lenticular nucleus varies in different cases from a discoloration and porosity to marked atrophy or disintegration. In extreme cases there is complete degeneration and cavity formation. Microscopically, there is destruction of the ganglion cells with marked proliferation of the glia and degenerative changes

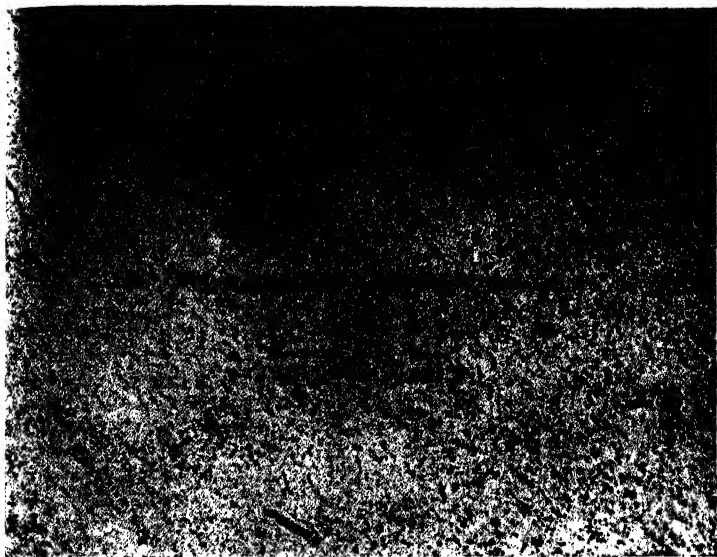


ILLUSTRATION 16.—PROGRESSIVE LENTICULAR DEGENERATION. Section from Left Lenticular Nucleus (see Illustration 15), Showing Marked Neuroglial Increase, Degenerated Nerve Cells in Various Stages with Tendency to the Formation of Satellitosis. (Courtesy of Drs. F. J. Farnell and A. M. Harrington, and the *Journal of Laboratory and Clinical Medicine*.)



ILLUSTRATION 17.—WESTPHAL-STRÜMPPELL'S PSEUDOSCLEROSIS. Various Forms of Atypical Large Glia Cells of Alzheimer. (a) Normal Glia Cell; All Other Cells are Pathologic. (Courtesy of Prof. A. Jakob.)

(see illustration 16), but without evidences of vascular occlusions or inflammatory lesions. In some of the cases there were also reported degenerative changes in the ansa lenticularis, relative atrophy of the corpus Luysii, partial atrophy of Forel's lenticular bundle and of the strio-Luysian fibers. In the chronic cases the lesions are generally more marked than in the acute cases, but this is not always so.

Pathologically Wilson's disease differs from Westphal⁵-Strümpell's⁶ pseudosclerosis in that in the former the degenerated areas of nerve tissue tend to soften; such is not the case in pseudosclerosis. In the latter there is a sclerosis which is diffuse, involving the strio-pallidum, optic thalamus, dentate nucleus, pons and cerebral cortex. The changes in the lenticular nuclei are minimal in pseudosclerosis, while in Wilson's disease the putamina are often totally destroyed. Alzheimer's giant glia cells (see illustration 17), so characteristic of pseudosclerosis, are never found in Wilson's disease. In a recent case reported by Greenfield, Poynton and Walshe⁷ in which the clinical symptoms were those of Wilson's disease, these authors found degeneration of the nerve cells in the putamina and caudate nuclei with an overgrowth of neuroglia; there were similar, though less marked, changes in the globus pallidus and red nucleus; Marchi preparations of the tracts leading from the putamen to the red nucleus and corpus Luysii, of the posterior longitudinal bundle, and of the superior cerebellar peduncles showed degeneration.

Owing to the great number of atypical cases reported (V. Economo, C. and O. Vogt, Bielschowsky, Henschen, Thomalla, Rausch and Schilder⁸ and others) as well as to the similarity of the histologic processes of Wilson's disease to some cases of met-encephalitis and even to arteriosclerotic disease of the brain, A. Jakob⁹ includes all these cases in one group which has been designated by Henschen as "spastic pseudosclerosis." Sachs¹⁰ is not inclined to regard the disease as a clinical entity. Strümpell⁶ believes that paralysis agitans, Wilson's disease and pseudosclerosis all belong to one group. Wilson¹¹ himself states that pseudosclerosis is a nosologic conception that has been utilized to include cases, which, owing to limited pathologic investigation, cannot be said to conform rigidly to the clinical and pathologic concept which he attaches to bilateral progressive lenticular degeneration in his attempt to establish a distinct clinical entity. "Whether or not Fleischer's¹² ring of corneal pigmentation can be regarded as a characteristic sign of pseudosclerosis," says Wilson, "remains to be seen."

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DYSTONIA MUSCULORUM DEFORMANS

Occurrence, p. 539—Symptomatology, p. 539—Treatment, p. 542—
 Prognosis, p. 543—Pathology, p. 544—Historical summary, p.
 545—Bibliography, p. 546.

Synonyms.—The disease has been described by different observers under different names, such as Dysbasia lordotica progressiva, Dystonia musculorum deformans (Oppenheim), Tonic torsion spasm (Ziehen), Progressive torsion spasm of children (Flatau-Sterling, Hunt), Ziehen-Oppenheim disease (Van Bernstein), Tortipelvis (Fraenkel), Dystonia lenticularis (Taylor) and Crampus syndrome (Förster).

Definition.—Dystonia is a symptom-complex characterized by alterations of tonus in certain single muscles or groups of muscles resulting in bizarre movements and constantly changing grotesque deformities.

Occurrence.—The disease was originally thought to affect mostly children of Russian-Jewish parentage, between the ages of eight and fourteen. With the increasing number of reports of cases it would seem that its occurrence is not confined to that race, nor does it always begin in childhood. It is sporadic rather than truly familial, although in a few instances more than one case has appeared in the same family. It occurs with greater frequency in females than in males. As our knowledge of the extrapyramidal motor disorders is increasing, the syndromes of dystonia, especially in association with, or as a sequela of epidemic encephalitis, is being more readily recognized clinically. This probably accounts for the greater number of cases reported in the literature during the last five years. The disease is apparently much more frequent than it was originally thought to be.

Symptomatology.—The affection is characterized by the presence of wave-like, more or less rhythmic, twisting, involuntary movements involving various groups of muscles. The movements usually begin in one upper extremity and gradually extend to the other limbs, but the

involvement is most marked in the muscles of the pelvic girdle. The movements are intensified during volitional acts and mental excitement. The affected muscles are alternately in a state of hypertonia and hypotonia. The constant twisting associated with the alternating changes in muscle tonus gives rise to most grotesque attitudes and deformities, which are especially striking around the pelvic girdle, the so-called "tortipelvis" (see illustration 19 A). Many cases show scoliosis, lordosis, wry-neck and curious deformities of the hands and feet ("semilunar foot"—Hunt). Owing to the tilting of the pelvis the gait is very char-



ILLUSTRATION 18.—PARAPLEGIC TYPE OF DYSTONIA MUSCULORUM DEFORMANS. Note the "Scissors Gait" Attitude of the Legs. (From the Neurologic Wards of the Montefiore Hospital.)

acteristic; the vertebral column is thrown forward producing a marked lordosis (see illustration 20 B), the lower extremities are everted, or inverted and flung about awkwardly in movements of hyperextension and hyperflexion. Some patients present a typical "dromedary" or "monkey" gait, while in others the tendency to hyperadduction produces a

"scissors-gait" effect (see illustration 18). The spasms as well as the deformities are diminished during rest, and cease entirely during sleep. Most patients learn to assume a definite fixed position, "posture of election" while standing, sitting or lying down, which gives them most relief from the spasms (see illustration 19 A and B). Occasionally, cer-



ILLUSTRATION 19.—A. ADVANCED CASE OF DYSTONIA MUSCULORUM DEFORMANS SHOWING INVOLVEMENT OF THE TRUNK, NECK MUSCLES AND ALL FOUR LIMBS. Note the Tilting of the Pelvis.

B. SAME PATIENT IN RECLINING POSTURE—"POSTURE OF ELECTION." Note the Hypotonicity of the Right Hand. When She Remained in This Attitude Her Movements Disappeared, and She Could Fall Asleep. (From the Neurologic Wards of Montefiore Hospital.)

tain voluntary movements of a limb would be made by the patient to influence or diminish the severe spasm, experience showing that such movements had a relaxing effect on the dystonic disturbance. The writer has observed two patients in whom the abnormal movements and deformity of the spine and pelvis largely disappeared when they "crept on all fours."

The fatigue and strain brought on by the spasms increase the rate of the pulse, and produce vasomotor disturbances such as flushing of the skin and hyperidrosis.

In uncomplicated cases the tendon reflexes are normal, but at times difficult to elicit owing to the deformities and contractures which may

exist. The superficial reflexes are normal and there are no pathologic reflexes. There are no paralyses, no muscular atrophies, and no changes in the electrical reactions. The sensations, sphincters and cranial nerves are all intact. There are no mental disturbances.

In a case described by Bernstein in 1912 there was a distinct dysarthria. In 1918 the writer also reported a case in which dysarthria and dysphagia were present. Since then Wimmer has reported a case in which speech, though not definitely dysarthric, was explosive; and Wechsler and Brock have also reported two cases with disturbances in speech. There is no reason why the speech muscles should not be involved in dystonia as well as the other muscles. Involvement of the muscles of articulation is not unusual in the dystonia syndromes following epidemic encephalitis. The writer has now under observation a 25 year old woman who had an attack of acute epidemic encephalitis five years ago which was followed by a dystonia involving the distal ends of the right upper and lower extremities, right sterno-cleido-mastoid and tongue. This patient also shows a parkinsonian tremor in the left hand. Wechsler and Brock also report a case of dystonia with a parkinsonian tremor of the right thumb and hand; their case apparently gave no history of a preceding encephalitis.

In dystonia all volitional movements are performed as if there were a conflict in action of the different muscle groups, a reversal of muscle tonus. There is an inability to dissociate harmoniously the essential muscle tonus elements of a simple movement, namely contraction of the agonists and antagonists. This, Hunt designates as the "paradoxical" or "reverse phenomenon" of dystonia, and he considers it diagnostic in the differentiation of this disease from spastic, hysterical and other forms of spasmodic contractures. The phenomenon is elicited in the following manner: The wrist joint being held in flexion by the patient, when he is requested to extend the wrist, he makes an attempt to do so, but instead of extension there are several involuntary flexions to a greater angle than there was originally, and after a perceptible lapse of time the desired extension is first executed. The same result may be obtained in any of the involved joints.

Treatment.—Therapy has had little or no effect. Before the organic nature of the affection was recognized various methods of treatment such as psychotherapy, electricity, hydrotherapy, metallotherapy, and the administration of extracts of the various glands of internal secretion were employed. In one of Fraenkel's cases, reëducation seemed to help for a time, as did the **intraspinal injection of magnesium sulphate**. Sedatives, such as **bromids, hyoscin, luminal** and even **morphin** are occasionally indicated to give these patients relief from the severe and exhausting spasms. In several cases at the Montefiore Hospital **general anesthesia with chloroform** was resorted to; relaxation occurred after the patients had been completely anesthetized. The patients stood the anesthesia very well, but upon its withdrawal and before full consciousness had been regained, the dystonic movements recurred and were just as severe as before the anesthesia. In several cases the application of a plaster-of-Paris cast to prevent contractures aggravated the condition. In one case in which the dystonia was limited to the lower limbs (paraplegic type), an epidural injection of physiologic salt solution was followed by a cessation of the movements for eight

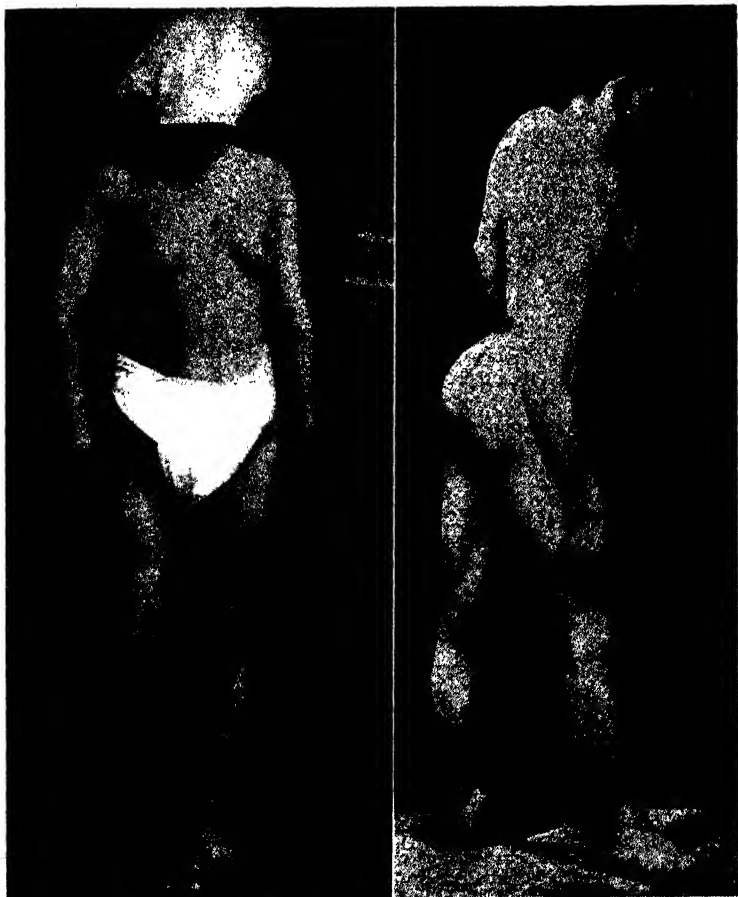


ILLUSTRATION 20.—DYSTONIA MUSCULORUM DEFORMANS. Hemiplegic Type. Right Hemidystonia More Marked in the Upper Extremity than in the Lower.

B. LORDOTIC ATTITUDE OF DYSTONIA. (Courtesy of Drs. Wechsler and Brock, and Archives of Neurology and Psychiatry.)

hours; in view of the fact that there was no paralysis of the limbs following this procedure, the writer is unable to explain this transient cessation of the dystonia. In still another case with isolated dystonia of one upper limb (monoplegic type), ramisection (Royle operation) of the cervical sympathetic chain on the side of the dystonia had no effect on the movements.

Prognosis.—The disease is chronic, progressive and incurable. The prognosis as to life is good. One of the writer's patients has had the disease for 16 years, and except for the dystonia which involves almost

every segment of his body, his health remains excellent. The patients, as a rule, die of some intercurrent disease. The progression of symptoms varies in different cases; one or two years may elapse between the onset of the dystonia in one limb and its extension to another limb. In one of our patients at the Montefiore Hospital the involvement of the limbs was hemilateral for a period of nine years (see illustration 20 A). Isolated dystonia is much more frequent in the post-encephalitic cases than in the "idiopathic" (?). The association of chorea, athetosis, and paralysis agitans tremor with dystonia is also much more frequent in the post-encephalitic cases. There are cases reported in the earlier literature in which improvement occurred spontaneously or after treatment, but such improvement was only temporary, to be followed by exacerbations, which were just as severe as the original attack. In the neurologic wards of the Mount Sinai Hospital, where we had opportunity to observe an unusually large number of cases of acute (?) dystonia associated with acute and subacute encephalitis, we were able to obtain remissions in several instances after we subjected the patients to progressively increasing doses of typhoid vaccine injected intravenously. As a rule the more severely these patients reacted to the vaccine the more certain and the longer were the remissions. None of the patients were permanently cured. The chronic "post-encephalitic" cases showed no remissions following several series of typhoid intravenous therapy, even though they reacted to the vaccine as severely as the acute cases.

Pathology.—Oppenheim believed the disease to be due to pathologic changes in the cells of the cortex which control muscle tonus. Fraenkel thought the disease to be due to faulty calcium metabolism. Studies of the calcium metabolism at the Montefiore Hospital on one of the patients afflicted with a severe type of the affection showed no abnormalities. Dana believed the disease to be a neurosis allied to general tic; Seelert also thought it was psychogenic in nature. Bonnhoeffer classed it among the choreas, and Bing among the athetoses. Jelliffe thought that the lesion involved some portion of the cerebello-thalamo-cortical arc, probably cortical to the red nucleus and possibly in the region of H' and H'' of Forel's field. Hunt, after an exhaustive clinical study of six cases, says that "the lower type of mechanism is at fault, one which is closely associated with the corpus striatum, but which is engaged in the regulation of tonus, especially in its relation to the reciprocal activities of the agonistic and antagonistic muscles, for this constitutes one of the essential factors in the motor disturbances of torsion spasm; but the preservation of facial expression and articulation is against a localization in the corpus striatum, for we know from many pathological studies that the face and articulation are both involved in this localization." Subsequent pathologic studies have shown the correctness of Hunt's hypothesis as far as localizing the lesions in the corpus striatum is concerned; it has also been shown clinically that the face and articulation may be involved in the disease, and that owing to the somatotopicality of the striatum corresponding to the various regions of the body, dystonia of the facial and of the speech muscles can be anatomically represented by lesions in that ganglion.

Only a few cases of dystonia have thus far come to necropsy. Ziehen's first case is said not to have shown any lesions in the nervous system. In 1918, Thomalla reported a case in which necropsy revealed a

"bilateral lesion, a total necrosis of the putamen and hepatic cirrhosis." He reports the case as one of "torsion spasm," and comments upon the similarity of the lesions to those found in "so-called Wilson's disease." Wimmer reports a case with a fairly typical clinical picture of dystonia in which he found cellular degeneration and neuroglial changes in the caudate and lenticular (putamen) nuclei, in the dentate nucleus of the cerebellum, in the thalamus, pons, cerebral cortex and spinal cord. The changes resembled those seen in pseudosclerosis and those found in the striatum were not more marked than elsewhere. According to Jakob, torsion spasm, like athetosis, is based on a pallidal lesion, both being dependent upon the relative functional capacity of the striatum; the latter must be at least partially intact in order that the pallidal athetoid hyperkinetic movements may develop. Hall also pointed out that the pathologic process is not limited to the lenticular nucleus, but involves the pons and cerebral cortex as well. He further states that degeneration of the liver may be found alike in progressive lenticular degeneration, pseudosclerosis and dystonia. Schneider is of the same opinion as to the hepatic cirrhosis in all these syndromes.

From all the pathologic evidence available it would seem that there is as yet no unanimity of opinion as regards the pathology of dystonia. All that one can say at this time is that Wilson's disease, pseudosclerosis and dystonia (according to Jakob, also perhaps some forms of athetosis) are variations in clinical expression of a pathologic process which is essentially the same, and that the predominating involvement is in the striatum. There is also no doubt that the so-called "idiopathic" variety of dystonia begins as a degenerative lesion *ab initio*, whereas the "post-encephalitic" variety begins as an inflammatory lesion, which goes on later to degeneration. It would also seem that whereas the maximal lesions in both varieties are in the striatum, there is a greater tendency to diffuse involvement of other systems in the "post-encephalitic" than in the idiopathic variety.

The etiology of the idiopathic variety remains wholly obscure.

Historical Summary.—At a meeting of the Berlin Psychiatric Society, held in December, 1910, Ziehen reported 5 cases of peculiar twisting spasms in children, which he called "torsion spasm." Three of these cases had been previously published by Von Schwalbe in 1908. In October, 1911, Oppenheim published the clinical histories of four similar cases, and outlined a symptom-complex to which he gave the name "Dystonia musculorum deformans." Numerous writers have since then called the disease "Oppenheim's disease." *This name is unfortunate because the symptom-complex of dystonia is apt to be confused with that of "myatonia congenita" which is also sometimes described in the literature as "Oppenheim's myatonia."* In his original paper on dystonia, Oppenheim stated that he had seen similar cases before that time, but that he did not know whether they were cases of hysteria, hysterical scoliosis, lordosis, or idiopathic bilateral athetosis. Since then numerous cases belonging to this group have been reported both abroad and in this country. Although the mechanism of the pathognomonic symptom of the disease, the "torsion spasm," still remains unknown, the organic basis of the condition has been fairly well established. Our clinical knowledge of the condition has been especially enhanced by the numerous instances of general and partial dystonia observed during the recent

epidemics of encephalitis. Both clinically and pathologically the condition bears such close resemblance to Wilson's disease, pseudosclerosis, double athetosis, and some forms of isolated tics, that, for the present, at least, it would be best to regard it merely as a variety of extrapyramidal disease whose motor disorder is predominatingly dystonic in nature, rather than as a distinct clinical entity.

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THE ATHETOSES

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Introduction.—In 1871 William Hammond of New York described a condition—"characterized by an inability to retain the fingers and toes in a fixed position into which they may be placed, and by their continuous motion, due to involuntary contractions that take place slowly, apparently as if with deliberation and great force"—which he called "athetosis" (α =without, $\tau\acute{\eta}\theta\alpha\iota$ =to place). The condition is also known as "mobile spasm."

This disorder of motility is not a disease *sui generis* but is a peculiar phenomenon observed in various organic diseases of the central and peripheral nervous system. It is much more frequent in diffuse cerebral disease (encephalitis, cortical sclerosis, porencephaly, etc.) than in focal lesions. Athetoid movements may occur in general paralysis, tabes, subacute combined sclerosis and in peripheral neuritis. Inasmuch, however, as in most of these conditions the mechanism of the movements is

based on ataxia, they are usually designated as "pseudoathetoid," limiting the term "athetosis" to movements due to diffuse and focal lesions of the brain which give rise to hemiplegia or diplegia. In the latter instance it is frequently combined with posthemiplegic contractures and nearly always affects only the fingers or toes.

Symptomatology.—Athetoid movements are slow, rhythmic, involuntary twisting spasms most marked in the fingers and wrists, and in the severer cases, in the forearm, elbow, shoulders and leg, where the most common movement is an involuntary hyperextension of the great toe. The movements are usually described as "writhing," "cramp-like," "vermicular," "serpentine," "ameboid," "like tentacles of polypi," etc. Typical athetoid movements consist of a separation and adduction movement combined with flexion and hyperextension; all fingers do not move at the same time or in the same direction; there may be alternate abduction and opposition of the thumb with flexion or extension of the wrist and pronation or supination of the forearm. The movements rarely involve the muscles of the face and neck (platysma) but when they do, they give rise to the most hideous grimacing and most bizarre contortions. The constant morbid excess of motion is frequently followed by a hypertrophy of the muscles and hypermobility of the joints in the involved segments.

As a rule, the movements disappear during sleep, although in severe cases they may be present even then; they are diminished in intensity during rest, and if the hands and feet are interlocked, they may cease entirely; the slightest physical or mental excitement intensifies them; attempts at active or passive movements of the affected, and occasionally even of the unaffected limb, are followed by a temporary cessation of the athetosis, only to be renewed with greater violence. The patients are sometimes able to restrain momentarily the abnormal movements, but they must make a supreme effort of the will to do so; enforced restraint is followed by unusually strong movements. The lower extremities are, as a rule, affected to a slighter degree than the upper; the spasm is extensor in type, and the foot tends to assume the position of talipes equinovarus.

In athetosis due to cerebral lesions the movements are usually associated with a spasticity or rigidity of the muscles in the segments involved. In some cases the athetoid movements may be combined with incoördination of voluntary movements varying from slight ataxia to violent jerky movements similar to those observed in multiple sclerosis with marked cerebellar involvement.

Athetosis per se has no effect on the reflexes; the condition of the latter, and the presence or absence of pathologic reflexes will depend on the localization of the lesion which gives rise to the athetosis.

Diagnosis.—The diagnosis of the condition with the characteristic, slow, gradual, rhythmic, worm-like movements involving most commonly the distal ends of a non-completely paralyzed limb, and only rarely the muscles of the face, tongue and neck, as a rule, presents no difficulties. Athetoid movements may resemble *choreiform* movements; close examination, however, will show that the latter begin abruptly, they are not rhythmic and do not involve continuously the same muscles or groups of muscles; in contrast to athetoid movements, during which there is a prolonged, though changing contraction of muscles with a tendency to

hypertonicity, choreiform movements are interrupted by periods of rest, varying in duration, during which the muscles become flaccid (atonic). In this connection it must be borne in mind that in practice one encounters motor affections which are transitions between chorea and athetosis, and which are strictly speaking neither purely athetoid nor purely choreiform; furthermore, both forms of movements may coëxist in the same case, giving rise to *choreoathetosis*.

There is no difficulty in distinguishing athetoid movements from *associated movements* when it is remembered that the latter are muscular contractions occurring, when movements are executed or attempted, in muscles not directly concerned in the movement attempted; often the corresponding muscles of the opposite side of the body, and of the face, participate in the associated movements.

Clinical Types.—Jakob recognizes three types of athetosis:



ILLUSTRATION 21.—CASE OF SYPHILITIC MUSCULAR RIGIDITY WITH ATHETOSIS OF THE LEFT ARM FOLLOWING AN APOLECTIC LESION (*foc*) IN THE PALLIDUM OF THE RIGHT SIDE. Status Cribatus in the Strio-Pallidum of Both Sides. Myelin Fiber Stain. Frontal Section. (Courtesy of Prof. A. Jakob.)

(1) **SYMPTOMATIC ATHETOSIS.**—This type is seen in cases of Wilson's disease, syphilis or arteriosclerosis of the brain, encephalitis, etc. According to Jakob athetosis in adults is found only in cases in which there are lesions in the globus pallidus; this nucleus, however, must be partially intact, otherwise this form of hyperkinesia cannot develop; if these lesions, however, enlarge or multiply, the athetosis ceases and muscular rigidity makes its appearance. The pallidum, like the striatum, has a somatotopical localization, the area representing the head lying orally, then the area for the upper extremity, then that for the trunk and finally that for the lower extremity. (See illustrations 22 and 23.)

(2) **ATHETOSIS OF EARLIEST CHILDHOOD.**—This form of athetosis is observed in Little's disease. The condition is congenital and is characterized by convulsions, muscular rigidity and athetosis; the intellect may remain unimpaired. The anatomic basis of this form is the so-called "status marmoratus of the striatum" (the Vogts) with the characteristic myelin fiber picture in this gray nucleus. It is due to a disturbance in development and is the only affection of the striatum giving rise to pure athetosis.



ILLUSTRATION 22.—CASE OF MUSCULAR RIGIDITY DUE TO CEREBRAL ARTERIO-SCLEROSIS WITH AN APOPLECTIFORM ATHETOSIS OF BOTH LOWER EXTREMITIES WITH TENDENCY TO TWISTING MOVEMENTS. Status Cribratus in the Strio-Pallidum of Both Sides; Large Symmetrical Foci (*foc*) in the Posterior Part of Both Ansa Lenticulares. Myelin Fiber Stain. Frontal Section. (Courtesy of Prof. A. Jakob.)

(3) **ATHETOSIS IN INFANTILE CEREBRAL PALSY.**—This variety is characterized by spastic hemiplegia, athetoid movements, feeble-mindedness and epileptic convulsions. These cases show anatomically the Bielschowsky type of cerebral hemiatrophy with degeneration of the third layer of the anterior central convolution in addition to a mild involvement of the striatum.

Modern writers do not agree as to whether "*athétose doublé*," or generalized bilateral athetosis of childhood and adolescence, belongs to Jakob's type 2 or to type 3. The lesions at the basis of *athétose doublé* are diffuse with special involvement of the globus pallidus (see illustration 23). Clinically this form appears as a concomitant of the serious spastic congenital diplegias, and is characterized by the presence of generalized, bilateral, spontaneous, symmetrical wriggling and twisting movements of the muscles of the face, neck and extremities. The abnormal movements are best elicited during attempts at voluntary movement; they are aggravated by the slightest mental excitement, and are generally, though not always, accompanied by hypertonicity of the involved

muscles. The peculiar contortions of the body with the grimacing of the face present a striking clinical picture not unlike that observed in Huntington's chorea. The involvement of the muscles of articulation interferes with speech; involvement of the muscles of the tongue and of the muscles of mastication interferes with the taking of food. The participa-



ILLUSTRATION 23.—A CASE OF ATHÉTOSE DOUBLE SHOWING THE STATUS DYS-MYELINATUS OF C. AND O. VOGT, IN THE PALLIDUM (*Pall*). These Cases Frequently Terminate in Hypertonic Akinesia. Myelin Fiber Stain. Frontal Section. (Courtesy of Prof. A. Jakob.)

tion of the diaphragm and of the muscles of respiration gives rise to a peculiar jerky and grunting form of breathing. It is important to remember that most pronounced forms of athétose double occur in cases with very little motor paralysis.

Treatment.—The treatment is purely symptomatic, and not very satisfactory. **Sedatives, systematic exercises, hydrotherapy, electricity,** and all sorts of **mechanical contrivances** for the purpose of **immobilization** have been employed, but the results have not been of sufficient benefit to recommend their use. **Surgical treatment** has been resorted to by Hammond, Spiller, Media and Bossi, Sir Horsley and others; the good results reported to have followed were in cases in which the movements were limited to groups of muscles or to one limb. In the bilateral form of the disease with diffuse involvement of the brain and associated with mental deficiency and epilepsy, surgery can promise very little hope.

Prognosis.—In the symptomatic form the prognosis of athetosis is that of the primary disease which gives rise to it. In the other forms the prognosis as to life is good. From the pathologic nature of the con-

dition it is obvious that the outlook for recovery is very poor, except perhaps in the cases in which syphilis is an etiologic factor. The disease lasts many years, death being due to some intercurrent complication.

Pathology.—The pathology has been discussed under the heading of clinical types.

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THE MYOCLONIAS

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Definition.—The myoclonias are characterized by the common symptom of rapid, involuntary, clonic contractions of single muscles or groups of muscles, occurring in paroxysms, and which produce no movements or the very slightest movements of the parts affected.

Symptomatology of Clinical Types.—PARAMYOCLONUS MULTIPLEX.—In 1881, Friedreich described a symptom-complex in which lightning-like, jerky contractions of the muscles is a predominating feature, and which he called "paramyoclonus multiplex." The involvement is symmetrical, affecting mainly the muscles of the extremities and the trunk, e.g., the latissimus dorsi, trapezii, gastrocnemii, quadriceps extensors, the pectorals, the recti abdominis, biceps humeri, triceps, etc. The facial muscles, as a rule, are not involved, but the larynx and diaphragm may sometimes be affected. The movements are confined to muscle fibers, bundles or individual muscles, and resemble fibrillary twitchings; at times the contractions cause the bulging of an entire muscle. These

twitchings occur at the rate of from 30-100 or more per minute, lasting for several minutes; they may occur every half hour or more often, depending upon the severity of the disease. The jerkings may be arrested by calling the patient's attention to them; they are aggravated by rest and distraction of the patient's attention; they are diminished or may cease entirely during sleep. The oscillations are irregular, violent ones alternating with weak ones. Although the involvement is symmetrical, the contractions are neither synchronous nor rhythmical. A tap upon a tendon, a touch of the skin or the mere exposure of it will start a jumping and vibrating of the muscles in all directions, and quite independently of one another. The twitchings after such stimulation will appear as if the muscles were shocked by a weak electrical current. No matter how violent the contractions are, they never produce movement in the limbs or joints to which the muscles are attached. L. P. Clarke¹ reports a case of this disease in a patient in whom the myoclonic contractions became most marked and uncontrollable when he turned around in bed to change his position. There are other similar cases reported in the literature.

The reflexes, superficial and deep, the myotatic and electrical irritability are normal; the cranial nerves and the sensibility are unaffected.

In the cases recorded, males seem to be more frequently affected than females; the symptoms begin at any time of life between puberty and the age of sixty, fifty being the most common age. The disease may occur spontaneously or after fright, trauma (Starr, Carrière, Bertrand), mental or physical exertion, and after infectious fevers (Raymond, Valebra, Sterling, Meynert). Many cases are said to occur in Italy after malaria. M. Lafforgue² reports a case in which pronounced myoclonic symptoms appeared in one of his patients on the tenth day after a severe attack of mumps which was complicated with orchitis. Remak saw a case following diphtheria.

The disease has been met with as a complication of lead poisoning (Leusche), of paresis (Grawitz, Williams), of anterior poliomyelitis (Bailey), of meningo-encephalitis (Clarke), and the writer has seen it complicate Friedreich's ataxia in twin brothers. Lenoble and Aubineau report a case complicated with glycosuria. (Autopsy in this case showed no lesions in the brain or cord.)

UNVERRICHT'S MYOCLONIA.—Unverricht has described a form of myoclonia which is characterized by its familial nature and its association with epilepsy. It has been known to affect successive generations. The twitchings in this special form involve the muscles of the tongue, pharynx and diaphragm. The epileptic seizures occur in the beginning at rare intervals, but become more frequent later in the disease; they occur especially at night. The frequency of the occurrence of the disease may be gathered from the following statistics: Shanahan met the disease 7 times in 2,150 cases of epilepsy, Williams 6 times in 794 cases, and Turner twice in 2,000 cases. In many of the cases the paramyoclonus antedates the epilepsy, and in many of the cases the epilepsy escapes recognition because the seizures may occur only during the night; in most of the cases, however, the epileptic seizures precede the myoclonia which usually develops at the age of ten. As the disease advances, the epilepsy diminishes and the myoclonia becomes more marked.

Rhein³ reports a case of familial myoclonus in a sister and brother in whom the typical myoclonia was not associated with any form of epilepsy.

In Unverricht's myoclonia the twitchings are intensified by the influence of emotion or after physical exertion. The tendon reflexes, myotatic and nerve irritability are increased. Lundborg has emphasized the tendency which patients afflicted with myoclonus have to develop a dementia similar to dementia precox, late in the disease.

LUNDBORG'S MYOCLONIA.—A variety of myoclonia, which seems to be quite prevalent in Sweden, was studied by Lundborg who found that during the early stages of the disease the patients had alternately good and bad days. He spoke of a "psychoclonic reaction" in which the symptoms are aggravated by emotion, a "psychotonic contraction" during which tonic contractions of certain muscles take place during embarrassment with consequent inhibition of movement, and of a "senso-clonic reaction" in which the myoclonia becomes very intense on the "bad days" after the slightest sensory stimulation.

Popoff⁴ reports a case under the name of MYOTONOCLOMIA in which a 19 year old male had developed, at thirteen, clonic contractions of the left wrist during voluntary movements; later the fingers became involved. Three years after this, tonic contractions developed in the muscles of the forearm, followed by tonic and clonic contractions of the muscles of the neck, and finally in the flexor muscles of the trunk; the spasms would last for two months after which they would disappear to return later. In addition to the myoclonic contractions, there was a tremor involving the entire body, which would increase when the patient attempted to straighten out his body. When sitting, lying down, or sleeping, the tonic and tremor would both disappear. The psyche was normal and there were no indications of organic nervous disease. The patient's father and younger brother showed similar myoclonic disturbances, but without involvement of the abdominal muscles.

Cases similar to those of Unverricht and Lundborg have been described by Buhner, Garnier, Verco, Putnam, Mott and others.

NYSTAGMUS-MYOCLONIA.—Lenoble-Aubineau⁵ described a rare and obscure disease which occurs almost exclusively in the Celtic races of Brittany and Great Britain. It is a hereditary, familial disease characterized by spasmodic twitchings of the external muscles of the eye and of the extremities, and tremor of the head. Exposure or percussion of the muscles intensifies the twitchings which can, to a certain degree, be voluntarily suppressed; the reflexes are exaggerated. Trophic and vasomotor disturbances are not uncommon. Stigmata of degeneration, such as deformed teeth, body and facial asymmetry, local hyperidrosis, circumscribed edema and lividity of the skin are often present. The disease, although incurable, does not seem to be progressive. No pathologic changes have been found in the cases examined.

N. S. Yawger,⁶ of Philadelphia, has recently reported several cases of familial head nystagmus in four generations associated with ocular nystagmus in a Russian-Jewish family. There were several stammerers in the family. Both sexes had the combined nystagmus and both transmitted it to their children. In none of the cases was there spontaneous nystagmus; in most of them the ocular movements, the speed of which was from 120–200, preceded the head movements, of which the patients were not conscious unless their attention was called to them. The head

movements were horizontal, in the same direction as the eye movements, and coördinated; the approximate frequency being 20 per minute. All the patients were mentally alert and had no other signs of organic nervous disease. Similar cases have been reported by Popper,⁷ Rosenfeld,⁸ Thompson,⁹ and Nettleship.¹⁰

MYOCLONIA AND EPIDEMIC ENCEPHALITIS.—Myoclonic movements are frequently observed in epidemic encephalitis, giving rise to the so-called "*myoclonic form of encephalitis*." In a few cases rhythmic movements of the distal portions of the extremities, abdominal or facial muscles appear after an attack of encephalitis; the myoclonic movements in these cases may be associated with parkinsonian and with choreiform or choreo-athetoid movements in the same, or in different segments of the body. These cases, however, are very rare. A more common form is the one designated by Hunt and others as "*acute infectious myoclonus multiplex*." In this form the disease begins with myoclonia. These patients present quite a uniform and rather striking clinical picture, which consists of severe headache, drowsiness followed by insomnia, lancinating pains, forceful, shock-like contractions of the abdominal muscles occurring at the rate of from 40 to 60 per minute, and a peculiar delirium. The delirium presents the characteristics of a toxic delirium with fleeting hallucinations, illusions and transitory delusions. These patients are extremely restless, apprehensive and confused. In the severe type in the later stage, there may be apathy and a tendency to stupor. The mental symptoms are more marked at night, and the delirium frequently assumes the characteristics of an occupational delirium. The lancinating pains are usually limited to the same segments as the myoclonic movements, giving rise to the radicular type of myoclonic encephalitis. In some of these cases the lancinating pains are frequently associated with hyperalgesia and hyperesthesia, not unlike that observed in posterior root irritation due to an infectious or neoplastic process, but without evidences of paralysis. The lancinating pains and myoclonic movements may, at the onset of the disease, begin in the trunk and extremities; at first they are local, but soon become generalized. As a rule, the shooting pains continue with diminishing severity over a period of one or two weeks, but occasionally they may persist with considerable intensity for three or even four weeks. The pains usually precede the characteristic muscle jerks which may be associated with muscle waves (myokymia) and fibrillary twitchings. In some cases, an interval of a week or more may elapse before the appearance of the myoclonus, myokymia and fibrillations. The muscular twitchings are bilateral, multiple and generalized, although a tendency to localization in certain regions of the body may also occur, and more especially in the trunk and abdominal muscles. The myoclonic contractions are quick and of short duration, involving individual muscles or groups of muscles, but not synergistic groups, so that the resulting locomotor effect is comparatively slight.

Hamill described a group of cases belonging to this type in all of which the movements were increased during sleep and during excitement, whereas voluntary movement had almost no effect. In Hamill's cases the movements involved the muscles used in forced respiration such as the sterno-cleido-mastoids, pectorals, latissimus dorsi and abdominal muscles.

Krebs describes rhythmic and arrhythmic myoclonic movements in

epidemic encephalitis; according to him, the arrhythmic type recalls the contractions of the paramyoclonus of Friedreich; the rhythmic type differs from the arrhythmic in that it moves the limbs. Hume, Natrass and Shaw¹¹ found myoclonic movements in 14 out of 20 cases of lethargic encephalitis; the movements were limited to the limbs in 9 cases, to the abdomen and back in 3, and to the face and jaw in 2. The cases reported in the literature as "epidemic hiccup" are probably examples of myoclonic encephalitis with special involvement of the diaphragm.

The prognosis of the myoclonic form of encephalitis is very grave. Most of the acute cases observed by the writer terminated fatally; the patients appeared toxic from the very onset of the disease and death was due to exhaustion. With a few exceptions, the cases that recovered showed a subsidence of the myoclonic movements that was simultaneous with that of the mental symptoms, the shooting pains persisting long after the myoclonic movements and the mental symptoms had disappeared. The similarity of the clinical picture as well as the fatal outcome of so many of the cases to that of "electric" or Dubini's chorea naturally raises the question whether the original cases reported by Dubini may not have been cases of myoclonic encephalitis.

Diagnosis.—The clonic, lightning-like, rapid, symmetrical, paroxysmal twitchings of the muscles of the trunk and extremities, without involving the face (except in the cases of myoclonic epidemic encephalitis), and producing no movement of the parts affected are sufficiently characteristic to establish the diagnosis of myoclonia.

Treatment.—Various sedatives, extracts of the different glands of internal secretion, galvanic electricity and hydrotherapy have been recommended. Psychotherapy is of service only in cases associated with hysteria. The writer obtained favorable results from very large doses of **chloral hydrate**, 25 to 40 grains (1.6–2.6 gm.); the results, however, were only temporary. **Hyoscin** and **bromids** seemed to have no effect. In the "encephalitic" cases the injection of typhoid vaccine (intravenously) was followed by no better results than after the administration of chloral. In several cases lumbar puncture was followed by a temporary cessation or alleviation of the myoclonic movements; such improvement was noted only in the cases in which the spinal fluid was under markedly increased pressure. Piticariu reports good results from the intravenous injections of the patient's spinal fluid. Once a week, he performed a lumbar puncture, and each time injected 10 c.c. of fluid intravenously. As many as seven such treatments were given.

Prognosis and Duration.—The prognosis as to recovery is not good. The cases which have been reported as cured were probably hysterical in nature. Remissions and relapses are not uncommon. The disease may last indefinitely; many patients have lived to be 70 years old. Death, as a rule, is due to deglutition pneumonia, or myoclonic spasm of the pharynx (rare involvement). In the older cases, death may be hastened by the onset of a gradual dementia and marasmus.

Pathology and Pathogenesis.—Heilig, on account of the facial involvement in some of the cases at the beginning of the disease, believes it to be a form of hysteria, tic, or chorea. Strümpell, Huchard, Fiessinger and others consider the disease identical with hysteria. Hoffman and Böttiger think the condition closely related to, or identical with chronic

chorea. Farge, Mettler, Williams, Lugaro-Soury and others consider myoclonic contractions a symptom of other diseases. Stadler¹² reports two cases in which in addition to the characteristic muscular contractions there was in each case a progressive bilateral atrophy affecting adjacent muscles, not in any particular groups, without fibrillary twitchings, reaction of degeneration, or sensory disturbances, and on account of these findings, he is inclined to consider the disease similar to myotonia (Thomsen's disease). Popoff also considers the disease myopathic in origin. Friedreich himself thought it is due to an irritation of the anterior horn cells, and Tutschaninow who reproduced the twitchings in dogs experimentally by injecting the spinal cord with carbolic acid, is inclined to agree with him.

Hoover and Vaulair believe the cause to be a hypersensitiveness of the receptor cells of the spinal cord. Hunt also thinks the disease is spinal in origin. Strasman contradicts this view. Lundborg thinks the contractions are produced by intestinal intoxication. V. Wagner-Jauregg observed in animals, deprived of their thyroid and parathyroid glands, spasmodic phenomena resembling myoclonia. Raymond thinks that myoclonia may be caused by hysteria, psychasthenia, or epilepsy and may accompany any organic disease of the nervous system. Murri, Seppilli, Patella, Massalongo, Clark and Prout consider the origin of the disease to be an irritation of the motor cortex. F. Schultze suggests that convulsive tic is a monoclonia, while general tic is a polyclonia; he believes Unverricht's myoclonia to be the same as chronic progressive chorea. Dana would regard myokymia a form of myoclonia. Oppenheim considers paramyoclonus multiplex an independent disease, and considers Unverricht's cases as being a special type of it, or a different disease, and he believes that in addition to these there are some obscure diseases in which myoclonic contractions may be symptoms of special importance. He calls attention to the difficulties encountered in the recognition of the hysterical myoclonias.

The results of anatomic examination have thus far simply added to the confusion which exists both as to the pathology as well as to the pathogenesis of the disease. In many cases, intense lymphocytic infiltration was found in the brain and cord. In a case of familial myoclonus epilepsy, Sioli¹³ found slight degeneration in the upper cervical cord, the so-called Hellweg triangular tract; the anterior horn cells were normal. The cerebral cortex was like that found in epilepsy, and in the cerebellum he found a large lipoid mass near the dentate nucleus, which extended into the white matter of the cerebellum and somewhat into the pons. Sioli does not know whether this lipoid mass was a chance finding, or whether it bears some relation to myoclonia. It may also be of interest to note that Hunt found a hypertrophy of the primary fibers in the muscles.

H. Roger is certain that these muscle changes are secondary. In some of the reported necropsies in cases of *epidemic encephalitis* showing myoclonic movements, lesions have been found in the pons, midbrain, locus niger, medulla oblongata and spinal cord. The variations of the pathologic findings in the reported cases, however, are so great as to leave the question unsettled. Hunt suggested that rhythmic myoclonia relates to a disturbance in the infrastriatal mechanism and that non-rhythmic myoclonia (paramyoclonus) relates to lesions in the medulla

and cord. This view is concurred in by H. Roger.¹⁴ In view of the fact that most cases of epidemic encephalitis show diffuse lesions scattered throughout the central and peripheral nervous system, it would seem almost impossible to localize definitely the lesions responsible for the myoclonia. On clinical grounds, one may perhaps be justified in attributing the myoclonic movements to an irritative lesion of the lower motor neuron.

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SPASMS

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LOCALIZED MUSCULAR SPASMS

General Considerations.—In general, it may be said that a "localized spasm" is a symptom and not a disease. It is defined by Brissaud as a reflex movement due to an irritation along the pathway of a periph-

eral reflex arc. The movements of a spasm are brusque, resembling the muscular contractions following electrical stimulation. The muscles involved correspond to the anatomic distribution of a nerve. If a portion of one nerve is involved, the spasm may be partial or fascicular, as in spasm complicating facial palsy; if the whole nerve is involved, the convulsive movement may be similar to the performance of a purposive act. In facial spasm, the forehead of the affected side is wrinkled by the contraction of the frontalis muscle, while the eye is closed by the contraction of the orbicularis palpebrarum, both of these muscles being innervated by the same nerve—the facial. Such combined movements cannot be produced at the same time voluntarily. This phenomenon is called by Babinski “paradoxical synergia.”

Facial Spasm.—SYNONYMS.—*Spasmus facialis*, *Convulsive tic*.

ETIOLOGY AND SYMPTOMATOLOGY.—Facial spasm may be the result of organic disease in the area of distribution of the sensory branch of the trigeminus, such as the cornea, conjunctiva, the nose, the teeth, the maxillary bones, the tonsils, etc. It is therefore often associated with “*tic douloureux*.” Any original physiological reflex movement in this distribution may gradually develop into a spasm involving the facial muscles.

Facial spasm begins with clonic contractions, which, as they advance, gain in rapidity, and at the height of the attack become tonic; as this subsides, the clonic movements reappear and remain until the attack is over; the entire cycle lasting about a minute. The twitchings may be observed on the return of power in previously paralyzed muscles, and they may be sufficiently severe to involve the muscles of the other side of the face. Partial facial spasm may be due to overstraining of individual muscles, such as the orbicularis palpebrarum in the course of certain occupations, commonly seen in watchmakers, microscopists, etc.; the spasms are then considered as symptoms of an “*occupation-neurosis*.”

A reflex facial spasm is sometimes noticed in diseases of the ear; it may follow a direct injury to a peripheral nerve, or injuries to the head, which have given rise to organic disease of the brain, or to a traumatic neurosis, i.e., merely from the shock of the injury. A good example of a facial spasm following irritation of a nerve trunk is found in recent literature in two cases of convulsive facial spasm reported by Cushing,¹ due to tumor of the cerebellopontine angle.

Affections in other parts of the body, such as disease of the generative organs, pregnancy, etc., may give rise to facial spasm. It may also be one of the complications of chorea, epilepsy, hysteria, migraine, the psychoses, etc. In many cases no pathological basis for the condition can be discovered, and it is then considered psychogenic; it has been known to develop after shock or great emotional excitement in neuro-pathic individuals, although direct inheritance is not common.

The most frequent form is hemispasm, confined to the orbicularis palpebrarum, although it may be bilateral, or diffuse and extend to the platysma, occipital and ear muscles. When it is limited to the orbicularis palpebrarum, and when the contractions are purely tonic in nature, it is called “*blepharospasm*,” but when clonic, it is called “*blepharoclonus*” or “*nictitation*.”

Mental excitement and physical overstrain aggravate the condition; distraction of the attention and suggestion temporarily inhibit it; it

may persist during sleep. A sudden examination of the eyes, or a tapping or stroking of the face will provoke it very promptly. Spontaneous remissions and exacerbations are common.

The *subjective symptoms* may be slight or severe, depending upon the intensity of the spasm; true facial spasm may be associated with, or provoked by pain in the trigeminal area. There is, as a rule, no interference with the voluntary movements of the implicated muscles. There are no vasomotor and no trophic disturbances.

TREATMENT.—If a **cause** can be found, it is to be **removed** whenever possible. In general, careful **dieting**, **change of climate**, **change of environment**, with a minimum amount of mental and physical work, and well regulated **hydrotherapeutic** measures are indicated. **Arsenic**, **bromids** and the **analgesics** may be resorted to. **Local subcutaneous injections of cocain and atropin** have been used with success as far as the spasm was concerned but the injections have been followed by paralysis of the local parts.

Within the last decade, **injections with alcohol** (70–80 per cent.) into the sheath of the facial nerve on the involved side have been used with success by various clinicians. A most careful technic is necessary. The method was originated by Schloesser²; a few drops of the alcohol are injected along the stylomastoid process as far as the base of the skull, slowly and continuously, until paralysis sets in; this paralysis of the muscles passes off gradually and the patient is relieved for from three to seven months, when the injection must be repeated.

In some of the more obstinate cases, when the supraorbital nerve has been found to be tender on pressure, and when during such pressure the spasm is relieved, resection of that nerve has been successful; simple section of the nerve, however, has no effect. **Stretching of the facial nerve** has resulted in some cases in a temporary improvement; the spasm, however, was followed by a permanent paralysis. Oppenheim prefers injections with alcohol to nerve stretching.

In recent cases, **counterirritation with plasters** behind the ear, the **galvanic current**, **diathermia**, **faradism**, **static electricity**, **D'Arsonval current**, etc., have been recommended.

Grossman, of New York City, has reported a large series of cases in which improvement followed **breathing and relaxation exercises**. Oppenheim also recommends **gymnastic exercises**. **Hypnotism** and **suggestion** have been found to be of benefit in the cases complicating hysteria.

PROGNOSIS.—The prognosis is not favorable; the condition is chronic, and may last months and years. Strümpell has seen cases in which remissions occurred during pregnancy. Occasionally, recovery takes place spontaneously or after treatment, but when the disease has lasted for a long period, the ultimate outlook for permanent recovery is very poor.

Spasms of the Muscles of Mastication.—The spasms are limited to the muscles supplied by the motor fifth cranial nerve; they may be tonic or clonic. When **tonic**, the jaws are clenched and the mouth cannot be opened actively or passively, thus resulting in a general failure of nutrition. This variety of spasm is known as “trismus,” and it occurs in tetanus and meningitis, less commonly in tetany and occasionally, temporarily, in epilepsy. Diseases of the pons and beginning acute bulbar palsy may be accompanied by trismus. It is rarely an isolated symp-

tom. It is usually of reflex origin due to disease in the temporo-maxillary joint, or to unerupted teeth. It may also occur in hysteria.

When the twitchings are *clonic* they appear as rhythmical movements of the lower jaw, usually in the vertical direction; the movements may be so violent as to cause the teeth to chatter as in rigor (chills). These are seen during general epileptic and hysterical convulsions, and during the onset of febrile diseases, infectious in nature. As an isolated symptom, except transiently in hysteria, it is very rare. The grinding of the teeth in nervous children and adults during sleep is considered a variety of this form of spasm. This form of spasm is also observed in trigeminal neuralgia, and as a reflex in diseases of the ear. Schwartz and Burnett have observed by otoscope the spasm in the tympanic membrane.

TREATMENT.—The treatment consists in the **removal of the cause** and in relieving the symptoms. **Psychotherapy** is indicated in the hysterical cases.

PROGNOSIS.—The prognosis, if not due to organic lesions, is favorable; it usually disappears within a few weeks or months.

Spasm of the Tongue.—**SYNONYMS.**—Glossal spasm, Spasm of the hypoglossal region.—The tongue may participate in the spasms of chorea, epilepsy and hysteria, but isolated spasms of the tongue are rare. When they do occur independently, they are tonic, clonic or mixed. They may interfere with chewing, swallowing and speech, the latter resulting in "aphthongia," a variety of stuttering.

ETIOLOGY.—The most common causes are emotional excitability, and reflex irritations from the throat, bad teeth and stomatitis. Strümpell mentions a case of spasm of the tongue in a glass blower. A neuropathic condition predisposes to the neurosis. "Tongue chewing" is frequently observed in infants during dentition, and in mentally defective children.

SYMPTOMATOLOGY.—The spasm is rarely continuous; it comes on in paroxysms every few days or weeks, occurring from 20 to 30 times a day. It is rarely seen during the night. An attack may last a few seconds or minutes, or may continue for hours; it does not always cease during sleep. B. Myers³ records an interesting case of "tongue chewing" which occurred in several members of the same family. The twitchings are sometimes preceded by paresthesias in the mouth and tongue.

TREATMENT.—The treatment consists in the employment of ordinary measures to maintain the patient's general health, the **removal of irritations**, the use of **sedatives**, the **application of galvanism** (the anode to the hypoglossal nerve), and **psychotherapy**. **Surgical measures** are hardly ever indicated. Lange stretched on one occasion the hypoglossal nerve, then resected it, and finally had to divide the geniohyoglossus muscle.

PROGNOSIS.—The condition may last for years, but recovery ultimately takes place.

Spasm of the Muscles Supplied by the Glossopharyngeal Nerve.—**SYNONYMS.**—Deglutition spasm, Pharyngismus.

OCCURRENCE.—This form seldom occurs spontaneously. It is usually seen in hysteria and organic nervous disease, such as bulbar gliosis, in the form of tabetic crises, or in tetanus.

Spasm of the Muscles of the Neck.—**SYNONYM.**—Torticollis.

ETIOLOGY.—Torticollis affects mostly individuals of a neuropathic or psychopathic constitution; it is frequently associated with the neuroses and psychoses, and the patients very often present stigmata of degeneration. Rheumatic torticollis and congenital wry-neck are not included in this form of spasm, nor is the spasm of the muscles of the neck due to trauma or reflex causes, such as diseased cervical vertebræ included under this heading. Peripheral irritation, even of the slightest degree, such as a tight collar, may in predisposed individuals give rise to spasm; spastic torticollis may also be seen in diseases of the ear or brain.

Chronic poisoning with alcohol, lead or mercury is an important etiological factor. The condition may follow malaria, influenza, typhoid fever, pneumonia, chill, trauma, exertion or overstraining of the eye muscles from an error of refraction. In infants, rickets is a common cause. Brissaud considers this form of spasm purely psychogenic, and Oppenheim fully agrees with him when he says "the primary cause in typical cases is the neuropathic or psychopathic diathesis, and given this constitution, a number of factors, mental excitement, trauma, and overstrain of the cervical and nuchal muscles may bring on the spasm." The same author believes that the muscles of the neck are, next to those of the face, most commonly implicated in the movements of expression, and that localized muscular spasm frequently owes its origin to the fact that some emotional process, instead of becoming fixed in the mind as an imperative recollection, immediately invades the motor sphere and discharges itself in the form of a motor action—a spasm.

SYMPTOMATOLOGY.—Spasm of the muscles of the neck may be unilateral or bilateral; it may be limited to one or to several muscles of the neck. The muscles most commonly affected are those supplied by the spinal accessory nerve; the superficial or deep muscles or both may be affected; the sterno-cleido-mastoid, however, is involved in most of the cases. The spasm may begin in one muscle and in the course of time spread to the others.

The spasm may be tonic or clonic, or both. Prolonged tonic spasm of the sterno-cleido-mastoid is, as a rule, due to rheumatism, diseased vertebræ, or a congenital shortening of the muscles. The localization of the spasm in different parts of the same or other muscles will result in corresponding faulty attitudes. In severe cases, and at the height of the attack, the spasm may extend to the muscles of the trunk or extremities, simulating almost an attack of grand mal. When the spasm consists of "nodding" movements, it is called "spasmus nutans" or "salaaming spasm"; this form is most commonly seen in children during dentition, and may occur only during the night, or when the child is asleep. Involvement of the inferior oblique muscle results in simple "rotation spasm" (tic rotatoire).

The spasms may be so slight as to be hardly noticeable, and of no inconvenience to the patient, or they may be so severe that speaking, eating and sleeping may be impossible, and in these severe cases the involved muscles may, as a result of overactivity, become hypertrophied. Emotion, self-observation and attempts to check the movements, aggravate them; physical and mental rest, and the distraction of the attention alleviate them. On lying down the spasms cease at once, to return as soon as the patient gets up. Brissaud has pointed out that some

patients are enabled to arrest the spasm by placing and holding a finger on the chin.

The spasms occur in paroxysms, but in some cases they may be continuous, at the rate of from 10 to 30 contractions per minute. Remissions and exacerbations are quite common. Except for the discomfort felt at the neck just before and during the spasms, there are no subjective or objective sensory disturbances, and there are no paralyses. In some cases, the spasms occur only during the night, but in the severe cases, even in the non-nocturnal ones, they may persist during sleep.

The psychic symptoms are those of the individual psychosis or neurosis, from which the patient may be suffering. Oppenheim saw a case in which the spasms of the muscles of the neck alternated with hallucinatory confusion. Gowers had a case in which the patient had melancholia for ten years before the onset of the spasm. In alcoholics, the spasms may be associated with delirium. Duchenne and others have recorded cases in which the condition was associated with writer's cramp.

DIAGNOSIS.—The condition is differentiated from *rheumatic torticollis* by the absence of pain and tenderness in the cervical muscles; from congenital torticollis, by the history of onset and the absence of changes in the cervical spine and in the muscles themselves. *Organic brain disease* will be excluded by the mode of onset, the presence of paralysis, and positive eye findings. At times it is almost impossible to differentiate this form of spasm from *general tic*, especially when the latter is not accompanied by "echolalia" (*see Tics*), or systematic, purposive movements; prolonged observation will in many cases make the diagnosis clear. It is well to bear in mind that general tic is usually preceded by a tic of the facial muscles, particularly the orbicularis palpebrarum. It is distinguished from *myoclonia* in that the latter is not limited to the muscles of the neck, and that the contractions are of lightning-like rapidity, and do not produce movements of the part. In *chorea*, the movements are irregular, the involvement is general, and the onset gradual. Its association with rheumatism and its manifestations is also diagnostic; but when the spasms coexist with chorea, the diagnosis may be impossible.

TREATMENT.—**General hygienic measures, light nutritious diet, a change of climate and occupation** with the removal of any discoverable causes are indicated. For the relief of the spasms, sedatives such as **bromids, gelsemium**, etc., are employed; the condition being a chronic one, the use of opiates and other habit-forming drugs is not advisable.

Orthopedic Measures.—The application of pads to press the head in the direction opposite to the spasm, not too tightly applied, may be of use.

Electricity.—If pressure points can be elicited, the anode should be applied to them; if not, the anode is placed over the spinal accessory nerve, and the cathode over the muscle. **Faradism** has also been employed with good results, and **massage** properly directed may be of benefit.

Gymnastics.—Systematic exercises in fixing the head may be used as adjuvants.

Isolation, Psychotherapy, etc.—**Isolation and psychotherapy**, in the form of hypnotism, are of value in cases in which mental symptoms are a prominent factor. The "inhibition treatment" is highly recommended

by Oppenheim and others. **Counterirritation**, in the form of **blisters** and the **cautery** to the **nape** of the **neck**, has resulted in marked improvement, and in some cases in permanent recovery.

Surgical Measures.—Section of the tendons, or section and stretching or resection of the spinal accessory nerve have been resorted to with varying results. Kocher and Quervain performed a total division of the tendons of the nape muscles, with more or less success in a number of cases. Brissaud is opposed to any surgical interference. At best, the improvement following surgical procedures is only temporary. No operation is to be performed unless the milder methods have been employed and have given no relief. Oppenheim ascribes the good results of surgical intervention to its effect as a counterirritant and its influence on the mind. Various forms of injections into the muscles have been employed, with results lauded by those who originated them.

PROGNOSIS.—The spasms may last for years or for a whole lifetime. They may attain a certain degree of severity and remain stationary. Usually they vary in intensity. Spontaneous recovery, or under treatment, may occur in the milder cases, especially when a removable cause can be discovered. In some few cases, the spasms may be very distressing and so severe that the afflicted individuals commit suicide.

PATHOLOGY.—The objective picture of the disease presents both an organic and functional coloring, so that many observers continue to consider the affection as a neurosis. The fact that so many cases yield to psychotherapy lends great support in favor of the psychogenic nature of the condition. Strümpell was the first one to suggest a striate syndrome for torticollis resulting in abnormal tonus, spasms, and motor disturbances (slowing of motion and even tremor). Cassirer⁴ believes that torticollis may be a variety of torsion spasm, and includes it in the amyostatic syndrome. Rosenow,⁵ working with microorganisms, obtained from infected areas of the nasopharynx, teeth and tonsils, injected intracerebrally, subdurally and intravenously into rabbits, found evidences of specific localizing properties manifested in the organism. The injected rabbits developed abnormal movements of the head, and, at necropsy, the brains revealed perivascular infiltration in the meninges, subcortex, basal nuclei, pons, medulla and cervical cord, corresponding respectively to particular groups of muscles affected during life. The writer has seen two cases of torticollis as a sequel of epidemic encephalitis. In both instances, the patients showed evidences of dystonia localized to the limbs associated with a parkinsonian tremor and classical torticollis. Were it not for the history and other evidences of basal ganglion disease the torticollis would have been considered in both cases as psychogenic in nature. In the absence of postmortem evidence as to the involvement of the basal ganglions and on purely clinical grounds, it would seem that there exist some cases of torticollis which may be due to inflammatory or degenerative disease of the basal ganglions, although the majority of cases observed in practice are probably psychogenic in origin.

Spasm of the Muscles of the Trunk and Extremities.—Spasm of the muscles of the trunk and extremities, not due to organic disease of the brain or cord, is rare. When such "idiopathic" spasm does exist, it may involve a single muscle, such as one of the rhomboidei, the levator anguli scapulæ, the deltoid, the latissimus dorsi, the pectorals, etc., on

either side, or the same muscle on both sides, or a group of muscles innervated by the same nerve or by the same pair of nerve roots. When the spasm involves the lower extremities, the muscles of the calves are most frequently affected, although the muscles of the hip, the extensors or flexors of the leg and foot, and even the cremasters and dartos have been found to be affected with spasm.

The spasms may be tonic or clonic, and may be due to endogenous or exogenous toxic conditions, infectious diseases, chill, trauma, mental or physical exhaustion, painful joints, amputation stumps and other reflex causes. The majority of those who suffer from these spasms are neurasthenic or psychasthenic individuals.

TREATMENT.—The spasm of the calf muscles is perhaps the one which is the most amenable to treatment. The treatment does not differ from that of the other varieties of spasm.

PROGNOSIS.—The prognosis as to permanent recovery is not very good; the spasms may last for months and years. They are exceedingly obstinate affections, and have a great tendency to recur.

Spasm of the Respiratory Muscles.—*Tonic spasm* of the diaphragm is occasionally met with in hysteria. The patients have a feeling of suffocation; the movements of the diaphragm cease during breathing, and this may be followed by acute pulmonary emphysema. Spasm of this muscle may also be met with in tetany and tetanus.

These cases are best treated by **cold douches** while in a hot bath, with **hot fomentations applied to the pit of the stomach, electricity to the phrenic nerve, and sedatives**. Obstinate cases may require **morphin hypodermatically**, and at times **general anesthesia with chloroform or ether** may be necessary to relieve the spasm.

Clonic spasm of the diaphragm is much more common than tonic spasm. The ordinary hiccough (singultus) is a good example of this form of spasm. It may, in severe cases, be so frequently and rapidly repeated as to interfere with speech, respiration and the taking of food. When it is a symptom of hysteria it is almost intractable. It may be due to reflex irritation from the gastro-intestinal or genito-urinary tract, or to undue emotion or excitement. Direct irritation of the phrenic nerve may also produce it; Strümpell mentions a case of mediastinal pericarditis characterized by this symptom. It is a grave symptom, commonly seen before death in organic brain disease, and in severe infections, especially of the peritoneum.

Spasmodic yawning (oscedo), spasmodic sneezing (ptarmus), spasmodic snoring (rhoncho-spasm) are seen from time to time in hysterical and other neurotic individuals. Spasmodic yawning may be an aura of an impending epileptic attack, and is sometimes seen in organic disease of the brain, particularly the cerebellum. Spasmodic attacks of coughing when not due to laryngeal crises, or disease of the external ear, throat, nose and abdomen are usually hysterical in nature.

Saltatory Reflex Spasm.—**SYNONYM.**—Static reflex spasm.

This form of spasm was first described by Bamberger. As soon as the patient puts his feet to the ground, he begins to dance and to jump around on account of the clonic contractions of the muscles of the calf. The dancing, as well as the spasms, disappear as soon as the patient resumes the recumbent position. In some cases, the mere touching of the soles of the feet may bring on the spasms. There may be no

other symptoms than those of general nervousness or hysteria, although Erlenmeyer and Kast report that they have found in these cases the superficial as well as the deep reflexes exaggerated. The disease occurs in both sexes and at any age; it may appear spontaneously, or may follow any of the infectious diseases. It has been known to occur as an occupation-neurosis in ballet dancers. Oppenheim considers it a symptom or a rare form of hysteria.

TREATMENT.—The *treatment* consists of **sedatives, electricity, wet packs and psychotherapy.**

PROGNOSIS.—The prognosis of this form of spasm is good; as a rule it does not last longer than a few weeks or months.

Camptocormia.—**HISTORY AND DEFINITION.**—Souques⁶ and Mme. Rosanoff-Saloff⁷ have described a form of “neuropathic pseudocontracture” of the muscles of the trunk, which they observed as one of the war neuroses in the last war. They called the condition “camptocormia” (formed from the words meaning “bending forward”).

ETIOLOGY AND SYMPTOMATOLOGY.—The condition is a functional one and is similar to normal bending forward of the body, except that the head is kept extended for the purpose of enlarging the field of vision. Except at the outset of the disease, walking is not interfered with, and the patients can readily bend down to pick up objects from the ground. They are unable to stand erect, and any attempt to straighten themselves out is followed by a prolonged tremor in the legs, but on lying down they can readily straighten and even hyperextend the back. The vertebral spines are neither tender nor painful, but the lumbar muscles may be tender to pressure. Neurological and x-ray examination is negative.

Almost all of the patients observed were victims of so-called “shell shock,” and although not actually wounded, they had been knocked down after an explosion with more or less loss of consciousness. The authors think that the condition is a neurosis to which neuropathic individuals are greatly predisposed.

TREATMENT.—For the treatment of the condition, Souques devised a **corset** which was applied when the patient’s back became straight on lying down; those individuals who could not straighten their backs, even when lying down, had to be anesthetized. Aside from this, **measures ordinarily employed in the treatment of the neuroses** were resorted to.

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TICS

Localized and general tic, p. 566—Definition, p. 566—Etiology, p. 566—Symptomatology, p. 566—Diagnosis, p. 567—Treatment, p. 568—Course and prognosis, p. 568—Historical summary, p. 568—References, p. 568.

LOCALIZED AND GENERAL TIC

Synonyms.—Tic general, *Maladie des tics*, *Erinnerungskraempfe* (Friedreich), *Maladie des tics impulsifs* (Marina-Jolly), *Myospasia impulsiva*.

Definition.—"Tic is a reflex, defensive, or voluntary movement, which has assumed an imperative character" (Oppenheim). It may consist of a single movement, an *isolated* tic; or of a number of movements of different muscles occurring at the same time, or in rapid succession; or it may be in the form of a *general* tic. There is usually a "plurality" of movements involving several muscles supplied by different nerves. Thus, we have a "sucking tic," a "snuffing tic," a "licking tic," a "biting tic," a "grinning tic," a "scratching tic," a "nodding tic," a "gulping tic," etc. Under this heading of tics and habit spasms may be included the neurodermatoses, such as "tic de l'epilation of Raymond," called by Besnier "tic trichomaniac." Others call this form of tic, "trichokryptomania," "trichotillomania" and "trichorrhexomania." These are all similar morbid states in which the affected individual has an ungovernable desire to pull one or more of his own hairs from the scalp, eyebrows, eyelashes, beard or mustache. There is a similar neurodermatosis, called "dermatothlasia," in which an apparently normal individual has a constant and uncontrollable desire to rub, scratch or irritate the skin in one or several parts of his body.

Curious tics are described by neurologists, who have had opportunities to observe some of the war neuroses. Mott reports the case of a pugilist who was terrified by the explosion of a bomb dropped by a Zeppelin, and who developed jerky purposive movements of the shoulder and head as if to avoid a blow, and facial grimaces such as a pugilist might assume in a fight.

Etiology.—Tic never appears before the age of seven or eight, the most common period of onset being at, or near puberty. The localized as well as the general form affects chiefly hereditarily neuropathic and psychopathic individuals, while direct hereditary transmission occurs only rarely. Trauma, infections, intoxications, "imitation" and mental strain are exciting factors.

The physiologic basis for this peculiar motor disorder must be sought in the cerebral cortex, but its anatomic substratum has not as yet been determined. In this connection it is of interest to note that Jakob¹ has recently reported a case in which the patient had a unilateral facial tic for more than a year; the onset of the tic was apoplecticiform; necropsy showed an arteriosclerotic cyst in the capsule of the caudate nucleus. Jakob, however, admits that the movement resembled that of an isolated choreiform movement.

Symptomatology.—A tic represents a regulated movement, in so far that it always attacks only definite groups of muscles. The move-

ment may, at first sight, appear voluntary and intentional; the patient shrugs his shoulders, throws his head back, strikes the table with his hand, scratches his nose or his beard, lifts up his collar, etc.: all of these movements being repeated again and again in a most stereotyped manner. The movements pass off as suddenly as they came; distracting the attention usually arrests them so that the patients can attend to their occupations or business. However violent a tic of the right arm or shoulder may be, the patient's handwriting shows no abnormality. As Patrick pointed out, when the impulse to "tic" can no longer be suppressed, the patient takes his pen from the paper, executes his tic and resumes the writing. In most tiquers the execution of the tic movement seems to be a necessity; it may, therefore, be interpreted as a release of tension under which the patient is laboring, and which is followed by a feeling of satisfaction. Attempts at too violent suppression of the tic aggravate it; the same may be said of physical and psychic excitement. In severe cases a tic may persist during sleep. Pain is never an accompaniment of tic.

If the motor disturbance affects muscular groups over the entire body, one speaks of "*maladie des tics*," or general tic. This is a very rare disease which usually develops in children between the ages of seven and fifteen, who have a hereditary predisposition. According to Guinon and Gilles de la Tourette the condition is characterized by the following symptoms: Twitching of the facial muscles, especially blinking of the eyes and rapid opening and shutting of the mouth; these are followed by twitching of the neck, shoulder, and arm muscles although any muscle may be involved. If the involvement is bilateral the movements are usually asymmetrical. In many cases the muscles of articulation, phonation and respiration may be involved. The movements are systematic and constantly repeated in the same way. Some patients suffer from obsessions and compulsory actions, such as counting the steps when walking, or counting the windows of the houses which they are passing. Others utter meaningless words or repeat the same words or sounds—*echolalia*; or use obscene words—*coprolalia*; or imitate movements—*echokinesis*. Individuals afflicted with a tic are usually absent-minded, and cannot concentrate their attention for any reasonable length of time. This is why children so affected are such poor scholars; they all lack full control of their will power.

Diagnosis.—The diagnosis of tic is often a difficult matter. Fully developed cases can hardly be mistaken. If the twitching is limited to the facial muscles, it may be mistaken for *facial spasm* (see p. 558). If the spasm extends to the muscles of the neck, the diagnosis is made probable, but it is only confirmed by the appearance of systematized tics. It is distinguished from *chorea* by the fact that in the latter the movements are continuous, purposeless, general and irregular. A choreic never performs the same movement twice in the same way; a tiquer always performs the same movement in the same stereotyped manner. The mental state of a choreic is entirely different from that of a tiquer; *echolalia* and *coprolalia* are never seen in *chorea*. In *hysteria* the twitchings appear suddenly after some excitement, and there are usually other evidences of hysteria, such as paresthesias, anesthetics, etc., and the movements are not as stereotyped as in tic. The differentiation from the

myoclonias may at times be almost impossible (*see* section on *Myoclonias*). In some of the *psychoses* there may be grimacing of the face and wriggling of the shoulders or limbs at the onset or at the height of the disease, but the history of the case and prolonged observation will clear up the diagnosis.

Treatment.—**Drugs** seem to have no effect, though **bromids** may alleviate the spasms. When the patients cannot rest on account of the movements, **chloral or chloroform inhalations** may be employed. Wagner (cited by Oppenheim) reports **thyroid extract** of value in a few cases. **Light hydrotherapy (wet packs)** and as much **rest and isolation** as possible are indicated. **Relaxation exercises** which keep the body and each part of it at rest, for short periods at the beginning, and gradually lengthened as the treatment goes on, and **exercises** with a view to strengthening the power of inhibition, have been originated by Meige and Feindel, and independently by Oppenheim² with good results. The former make their patients drill before a mirror and call the exercise “psychomotor discipline.” Pitres and Cruchet, Grossman and others recommend carefully directed breathing exercises. Clarke,³ and Oberndorf report cases of tic cured by **psychoanalysis**. On the whole, it may be said that true tic is more often benefited by **treatment properly directed**, such as **exercises of “control,”** and by **due attention to the psychic elements of the case** than by any other means.

Course and Prognosis.—The condition is a chronic and progressive one, but complete recovery may take place in cases of even long duration.

Historical Summary.—The term “tic” is adopted from the French. The condition was recognized by Friedreich and also by Charcot and his pupils, particularly Gilles de la Tourette⁴ and Guinon.⁵ Further notable contributions on this subject came from Brissaud and his pupils, Meige and Feindel. The French school defined tics as “physiological acts, originally purposeful, but which have become acts apparently purposeless and meaningless.” The French investigators considered tics as psychoneurotic manifestations in individuals with infantile minds in regard to their emotional reactions.

When the freudian school began the study of the psychoneuroses by psychoanalysis, the mechanism of tic naturally attracted its attention, and it evolved the theory that a tic generally represents a purpose, that the purpose had been suppressed, and that the apparently senseless movement, when resumed, was a defense compromise which gave relief to the patient. L. Pierce Clarke³ from a psychoanalytic study of three stubborn cases of “mental torticollis” is inclined to emphasize the auto-erotic gratification unconsciously afforded the tiquer by the tic, and to regard the aspect of the tic as a defense compromise to be of secondary importance.

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OCCUPATION SPASMS

General considerations, p. 569—Etiology, p. 569—Symptomatology, p. 569—Diagnosis, p. 570—Treatment, p. 570—Course and prognosis, p. 570—Pathology, p. 570—Bibliography, p. 570.

Synonyms.—Occupation neuroses, occupation cramps, coördinated occupation neuroses, occupational dyskinesias.

General Considerations.—Occupation spasms are characterized by cramp-like contractions in the muscles, appearing only during certain definite complicated movements which have been acquired by habit or practice, whereas all other actions performed by the same muscles are normal.

In tailors and seamstresses, the spasms affect the muscles of the thumb and forefinger. Telegraphers, pianists, cigar makers, shoemakers, blacksmiths, automobilists, milkers, barbers, drummers, tennis players, leather dressers, and others may be similarly affected. The lips may be involved in trumpet players, the vocal cords in singers, the eyes in watchmakers, microscopists and miners ("miner's nystagmus"), and the calf muscles in dancers.

Writer's cramp (graphospasm) is the most common neurosis of this group. The onset is gradual; the patient first becomes tired, losing the usual control of his pen; he does not write as rapidly or as smoothly. After a time, owing to the spasm of the muscles, he finds that he grips the penholder too tightly, and that the writing appears irregular and incomplete. The muscles involved are the interossei, the thenar and hypothenar, the lumbricales, the flexors and extensors of the fingers and wrist, and the pronators and supinators. The more he thinks about the condition, the worse it becomes. Some patients have a simple weakness in writing; the muscles retain their normal power except during writing, when they seem to be paralyzed (?) (paralytic form), so that the process of writing becomes absolutely impossible.

According to Duchenne these disorders are classified as motor and sensory, including under the former the akinetic, hyperkinetic (classical spasm or cramp) and ataxic (tremors, choreiform and ataxic movement) types. The purely sensory type is extremely rare, but, on the other hand, sensory disorders of some kind, such as anesthetics, hyperesthesias, paresthesias and "neuralgias" are frequently encountered as complicating features in the more essentially motor types.

Etiology.—Occupation cramps are most commonly seen in adult males of neurotic make-up, so that more than one member of the same family may be affected. Physical and mental strain, bad posture during work, badly constructed and faultily held tools or instruments are exciting causes. Unusual attention to the work on hand, and anxiety as to the possible inability to go on with the usual occupation aggravate the neurosis and increase the spasms. It is a curious fact that, in violinists' cramps, the left hand may be involved as well as the right. In only a few cases has the condition followed injuries to the hand or local organic disease. There can be no doubt, however, that it may be produced in a reflex way by such conditions, especially when they are painful, and possibly also by a neuritis (Oppenheim).

Symptomatology.—The subjects of this disease frequently complain of paresthesias, local weakness, tremor and pains in the affected limb,

but objective examination fails to reveal any evidences of an organic motor or sensory disorder. There is no genuine tremor or ataxia. Owing to the associated neurasthenia the tendon reflexes are usually increased with an exaggeration of the mechanical excitability of the muscles. Local cyanosis, hyperidrosis, ischemia, and other vasomotor disturbances are not uncommon.

Diagnosis.—The disease must not be confused with organic nervous disease, such as tabes, disseminated sclerosis, hemiplegia, paralysis agitans and other conditions in which difficulty or inability to carry out complex movements is a prominent symptom. Writer's cramp has been wrongly diagnosed when the patient was suffering from agraphia, due to a tumor of the brain. A careful neurological examination with the finding of paralyses, sensory disturbances, electrical changes in the muscles or nerves, will make the diagnosis clear. The differentiation may at times be difficult when a hysterical or neurasthenic patient is, on account of tremor, unable to write.

Treatment.—The treatment to be carried out is with a view of improving the patient's general condition by **proper nourishment, hydrotherapy, change of environment** and other similar measures. Drugs seem to have little or no effect. Whenever possible, **absolute avoidance of the occupation** which brings on the neurosis is advisable. If for economic reasons this cannot be carried out, the patient is to be instructed to assume a **correct posture** while at work, and he is to be provided with **properly constructed tools or instruments**. **Massage, gymnastic exercises, and Swedish movements** are indicated. In writer's cramp, special penholders have been constructed which are held between the index and middle fingers (Zabludowski); a Nussbaum bracelet which makes the flexors and adductors superfluous during writing may be employed with benefit; in some cases the use of the typewriter may be necessary.

Course and Prognosis.—The neurosis may last throughout the patient's life. Some patients learn to write with the left hand, but sooner or later this hand also becomes affected. Recovery may take place, but relapses are very common. As in all the neuroses, the longer the duration the worse the outlook for recovery.

Pathology.—No lesions have been found to explain the malady. Various hypotheses have been advanced to explain its pathogenesis. Some authors favor the theory of perverted vasomotor habit dependent presumably on basic autonomic imbalance or dysfunction; others regard it as a variety of hysteria, and still others as an exhaustion neurosis.

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THE MYOTONIAS

THE MYOTONIAS

General considerations, p. 571—Myotonia congenita, p. 572—Etiology, p. 572—Symptomatology, p. 573—Myotatic irritability, p. 573—Electrical reactions, p. 574—Diagnosis, p. 574—Myotonia atrophica, p. 574—Paramyotonia congenita, p. 578—Treatment, p. 578—Prognosis, p. 579—Pathology and pathogenesis, p. 579—Historical summary, p. 579—References, p. 579.

General Considerations.—The myotonias are characterized by the presence of a peculiar motor disorder—*myotonia*—during which there occurs an inhibition of voluntary movements, due to a temporary cramp, or spasm of the muscles involved (see illustrations 28, 29 and 30). There is a persistence of contraction and partial or total inability to relax the muscles after the contraction. Whenever the patient begins to perform a brisk voluntary movement, the particular muscles participating in the intended movement are thrown into a state of tonic contraction which does not relax but passes off gradually, the muscles slowly becoming supple until, after a lapse of time, he can perform the movement desired. But if he stops and starts again, or if he tries to hurry his originally intended speed, this tonic stiffness and slowness of relaxation reappear, and have again to be, as it were, “worked off.” The period necessary for the spasm to yield may vary from 5 to 30 seconds. The myotonia (designated by some as “myotonus”) is at its height during the *second* and not during the first movement. For instance, if the patient wishes to carry out a quick and forceful movement with the thumb, he may succeed fairly well with the first adduction of the thumb, but the succeeding abduction necessary to carry out or to complete the intended movement will meet with very strong resistance, which can only be overcome upon the further attempt at repetition of the movement, which then becomes freer and freer until the tension is completely relaxed, and the movement eventually carried out in a perfectly normal manner.

Numerous symptom-complexes in which myotonus is a prominent clinical feature are described in the literature. Myotonia has been observed in diseases of the nervous system involving the cerebrum, cerebellum, spinal cord and peripheral nerves. The relationship of the fronto-ponto-cerebellar system to various myotonic phenomena has been emphasized by Kleist¹ who mentions the occurrence of myotonia in myelitis and syringomyelia with the typical mechanical and electrical myotonic response (see below) of the peripheral localization. There are other central types of myotonia, however, in which the mechanical and electrical responses are lacking and in which the myotonia is not diminished by repetition of the movement. The same author has also observed this type of myotonia in Friedreich's ataxia and Marie's cerebellar ataxia. Kinnier Wilson and Walshe² have also made the important observation that neoplasms of the frontal lobe anterior to the motor area may produce myotonia, a peculiar feature of which is the limitation of the disturbance to voluntary movements. Bumke³ also described a familial disease characterized by the occurrence of myotonia with both voluntary and automatic movements.

The mode and time of onset and distribution of the myotonia, as well as the course of the disease have led to the description of different types. Jacoby,⁴ for instance, makes a sharp distinction between the

congenital, acquired, and transitory forms. He reports a case of myotonia which developed after an attack of typhoid fever, and another case following trauma and overstrain. Beco⁶ saw a case develop in a young man with no history of the disease in his family, one year after he had suffered from two abscesses in his foot. Acquired forms of myotonia are also described by Ascenzi,⁶ Talma,⁷ Schott, Dereum,⁸ and others. It seems that in most of these cases, trauma or infection or both played a prominent etiologic rôle, and in many of them the myotonia was transient and curable.

Quensel⁹ records a case which he calls *pseudo-myotonia hemiplegica* in a man of 45 who had a peduncular hemorrhage, with disability on the left side, and in whom myotonic contractions always followed voluntary movements. In the writer's opinion this was probably a case of "symptomatic" myotonia due to a central lesion, similar to the cases that Stecker and Barkman¹⁰ have in mind, when they attempt to attribute myotonia to disease of the lenticular nucleus.

In the present state of our knowledge the most plausible approach to the entire problem of myotonia, at least as far as its nosology is concerned, would be to regard myotonia as a symptom-complex which may manifest itself in the form of a disease sui generis, or as a symptom, or complication of organic central or peripheral nervous disease. The writer, therefore, considers all the above cited cases as "symptomatic" myotonias, and *myotonia congenita* (Thomsen's disease), *myotonia atrophica*, and *paramyotonia congenita* (Eulenberg²¹) as distinct clinical entities, with the first as the purest form of the group.

MYOTONIA CONGENITA (THOMSEN'S DISEASE)

Synonyms.—Thomsen's disease, myotonia hereditaria (Nissen).

Definition.—Myotonia congenita is a hereditary disease characterized by cramps in the muscles at the beginning of voluntary movements, which disappear on repetition of the movements.

Etiology.—PREDISPOSING CAUSES.—*Sex.*—The disease has a preference for males, beginning in typical cases usually in early youth or about the time of puberty.

Mental Shock and Physical Overstrain.—Mental excitement and physical overstrain are said to hasten the onset of the disease; this is probably due to the fact that the shock or fatigue has aggravated the condition which had already been in existence, but the symptoms were so slight that they escaped detection. Individuals afflicted with myotonia congenita are generally unusually well developed, full-blooded and well nourished; their muscles are unusually large, but their actual strength markedly diminished.

Hereditary Influences.—The malady is hereditary, and usually affects several members of the same family. In accord with Rudin, Nissen¹¹ found that the affection followed the Mendelian law. Five chief factors were elicited: (1) direct heredity; (2) transmission only through dominants; (3) once free, always free; (4) a dominant marrying a healthy person resulted in one-half dominance and one-half unaffected; (5) the majority of members in large families were affected. According to Nissen, if two recessives marry, no dominance will appear but the condition will remain recessive. Because the disease is not always con-

genital, Gowers suggests that the designation "transient myotonia" would be a more appropriate one.

Distribution.—Up to 1914 H. Koch collected more than one hundred cases. The disease has been met with in Germany, Austria, Italy, France, Russia, Sweden, England and America. The original reports seemed to have been more numerous in Germany and in the Scandinavian countries than in other localities. The disease is considered to be a rare one, but this is probably due to the fact that some of the cases are so slight that they do not attract attention until perhaps later in life, especially in military countries, when the male members of the population are called upon to perform military service.

Symptomatology.—The first and most characteristic symptom is the stiffness in the muscles, the *myotonia* (or *myotonus*, see under General Considerations, p. 571).

The myotonus is very marked when the patient is suddenly called upon to make a rapid and forceful complex act, such as attempting to rise and walk after he has been sitting in a chair for a time. Going up and down stairs, with the necessary alternate relaxation and contraction of the muscles of the leg, is very difficult. These patients must be very careful on crossing streets busy with vehicular traffic, or while getting on and off cars; in the latter case it may be impossible for them to release the hand rail with sufficient rapidity to avoid being thrown against the car when getting on, or to the street when getting off.

Excitement, fright, dampness and cold aggravate the myotonus; mental rest, warmth and the ingestion of small amounts of alcohol diminish it. The condition is worse when the patient thinks he is being observed. The more prolonged the rest, the severer the spasm; when it has once passed off the patient can carry out the most complicated and most delicate acts without the slightest difficulty or fatigue.

The affection may involve all the muscles of the body, but it is most frequent in the limbs, and least in the muscles of the face and jaw. The ciliary muscles may be involved so that the patients find it difficult to accommodate for objects at different distances. In one of Charcot's patients, the eyeballs, on looking upward, remained in that position for a considerable length of time. Involvement of the tongue is not uncommon, but rarely are the muscles of the throat and respiration affected. The heart muscle is said to have been involved in some cases; Boot¹² reports a case of this disease in a boy in whom the heart sounds were unusually loud. The arms may be free and the legs affected, or vice versa. There are cases in which the disease is limited to certain groups of muscles. Oppenheim, Gaupp, Schott, Curschmann and others have described such atypical cases of partial myotonia, but most of them were associated with muscular atrophies. Sometimes the spasm is more marked on one side of the body than on the other. According to Thomsen, the more the muscles are used the less severe is the spasm.

The *myotatic irritability* of the muscles is markedly increased. Percussion of a muscle promptly gives rise to a slow but persistent tonic contraction; the belly of the muscle protrudes like a tumor, or a deep depression or furrow results where it has been struck. These phenomena are most noticeable in the tongue, the thenar and hypothenar eminences and the gastrocnemii. The slightest mechanical stimulation

will have the same effect; the muscular contractions may last from 5 to 30 seconds, instead of relaxing promptly as in health.

Mechanical or electrical stimulation of the nerve trunks elicits no peculiar changes in its excitability, so that no myotonus follows *indirect* electrical stimulation. The change in muscle irritability following *direct* electrical stimulation is a prominent and most characteristic symptom of the disease. This is known as Erb's myotonic reaction, "MyR."

The electrical excitability of the muscles is markedly increased for both currents. To galvanism $KCC = ACC$ (in health $KCC > ACC$); the contractions are *sluggish* and continue long after the stimulus has ceased. The stable galvanic current produces a rhythmic undulation of the muscle and the wave of contraction passes from the cathode to the anode; strong currents are necessary to produce this phenomenon. With the usual faradic (direct) stimulation, employing strong currents, there is a somewhat slow contraction with a long prolongation (2 to 20 seconds or more); single opening shocks of the slightest strength cause only normal quick contractions; with strong, stable faradization, oscillating muscular waves are occasionally seen. There is practically no change of reaction to static electricity.

There are no symptoms referable to the nervous system. The cranial nerves, sensation and sphincters are unaffected; the superficial reflexes remain unaltered; the tendon reflexes are active, but they have been reported diminished or easily exhausted. There are no trophic and no vasomotor disturbances.

Patients with this disease are said to take anesthetics very badly.

Most individuals afflicted with the disease assume a peculiar mental attitude in that they are reluctant to admit that they suffer from any muscular disorder. Jendrassik¹³ mentions the fact that "the patients seek to conceal their infirmity by all sorts of devices." Thomsen¹⁴ himself must have been thoroughly aware of his own psychic attitude when he said: "There is an active psychic factor involved, namely the fear of ridicule. As soon as the feeling arrives that those about will discern and observe the disability, the symptoms become aggravated in the highest degree." When psychoses do occur they are probably coincidental, although Rosett¹⁵ describes one family afflicted with the disease, in which the second generation consisted of two groups: one a myotonic group, and another group all members of which showed evidences of a border-line psychosis. It is also of interest to note that a number of Rosett's patients suffered with muscular pains, although the prevailing opinion seems to be that the disease is not painful but a source of great annoyance and, as Thomsen says, "it casts a shadow over the lives of the sufferers."

Diagnosis.—When the clinical features of the disease with the characteristic electrical changes in the muscles are taken into consideration, the diagnosis presents no difficulties. E. W. Taylor¹⁶ records the case of an Italian shoemaker, 24 years of age, in whom the disease was considered for a long period to be hysteria, because the spasms occurred in paroxysms between which the patient showed no signs of myotonia, and because there were no other members of the family afflicted with a similar condition.

MYOTONIA ATROPHICA.—This form of myotonia was described by

Rossolimo¹⁷ in 1902. It is also known as "amyotrophic myotonia" and as "dystrophia myotonica." The essential characteristics of the disease are the myotonia and the muscular atrophy. In view of the presence of muscular atrophy, some authors include the disease among the myopathies (muscular dystrophies), but inasmuch as, according to Pelz,¹⁸ 12 per cent. of all cases of myotonia congenita show muscular atrophy, other authors regard it merely an atypical form of myotonia congenita, while still others regard it a distinct clinical entity.



ILLUSTRATION 24.—MYOPATHIC FACIES (LONG, THIN AND TAPERING). Note Expression of Face, Bilateral Ptosis, Right Greater Than Left. (J. Neurol. and Psychopathology.) From the Neurologic Service of the Montefiore Hospital.

ILLUSTRATION 25.—MYOPATHIC FACIES. Note Similarity in Length of Face and Chin to that of Patient's Sister in Illustration 24. (Jour. Neurol. and Psychopathology.) From the Neurologic Service of the Montefiore Hospital.

In contrast to myotonia congenita, myotonia atrophica does not appear until the second or third decade of life. Males are more often affected than females. The myotonia generally precedes the atrophy and, as the disease advances, marked atrophies develop in some muscles, associated with myotonia (either in these or other muscles).

The muscles most commonly atrophied are: the orbicularis palpebrarum, the orbicularis oris, the temporal muscles, the sterno-cleido-mastoids, the vasti, and the anterior tibial extensors. In some cases the atrophy involves the same muscles as in the typical Erb's type of dystrophy, or as in the Aran-Duchenne type of progressive muscular atrophy (see illustrations 24, 25, 26 and 27).

Kennedy¹⁹ has pointed out a peculiar feature in this form of myo-

tonia, viz., that there is an extraordinary similarity in the appearance of the patients; "they all seem to look as if they belonged to the same family—like brothers and sisters."



ILLUSTRATION 26.—LATERAL VIEW, SHOWING HATCHET-LIKE FACIES OF SISTER. (J. Neurol. and Psychopathology.)

ILLUSTRATION 27.—LATERAL VIEW, SHOWING HATCHET-LIKE FACIES OF BROTHER. (J. Neurol. and Psychopathology.)

In typical cases the usual myotonic electrical reaction is modified in that the atrophied muscles show the usual diminished excitability to faradism, and to a lesser extent to galvanism. Occasionally the presence of a *myasthenic* reaction may be demonstrated. Some cases show in localized muscle groups a modified myotonic response—a slow contraction and slow relaxation to both faradic and galvanic stimulation. A wave of contraction may at times be elicited with a very strong current passing from cathode to anode. There is no reaction of degeneration present, except in the muscles that have completely atrophied.

The condition of the reflexes depends upon the degree of wasting in the atrophied muscles. In pure cases of the disease there are no fibrillary twitchings, no sensory changes, and no symptoms referable to the cranial nerves or to the sphincters. Hirschfeld recently reported a case in which, in addition to the symptoms of myotonia atrophica, the patient had atrophy of the shoulder-girdle and periods of bradycardia. After spinal puncture the pulse dropped to 40 per minute and continued so for a week. Under atropin it rose to 64. The cardiogram showed a slowing of the entire cycle, particularly the ventricular systole. The patient gave a history of previous similar attacks without cause.

Another characteristic feature of the disease is its association with congenital cataract. This association is too frequent to be a mere coincidence. It is rather suggestive that both myotonia and cataract in the same individual are evidences of an abiotrophy. Greenfield²⁰ noticed

that there were some families in which some members showed cataract without myotonia, others, myotonia without cataract, and still others, myotonia and cataract. The disease has also been found to be associated with other evidences of abiotrophy, such as the loss of hair, genital hypoplasia, and absent reflexes with tabiform degeneration of the cord. In the male, the most frequent of these extramuscular symptoms is baldness, with atrophy of the testicles next in frequency. There is no doubt, however, that the sexual functions are affected sooner or later in a very large number of cases in both sexes, and it is therefore reasonable to believe that the almost constant loss of libido and of sexual power is due to changes occurring in these organs. In some patients the external genitals

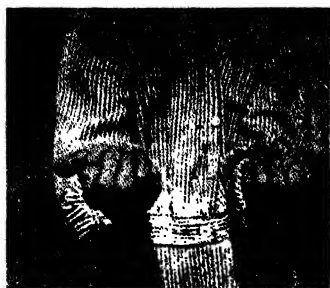


ILLUSTRATION 28.—TYPICAL MYOTONIC REACTION AS SHOWN IN BOTH HANDS, Patient Attempting Initial Movement in Opening Closed Fist. (J. Neurol. and Psychopathology.)

ILLUSTRATION 29.—SAME AS ILLUSTRATION 28, IN THE CASE OF THE BROTHER. (J. Neurol. and Psychopathology.)

are infantile. Celibacy and childless marriages are common. Loss of body weight, acrocyanosis, increased secretion of tears, and marked asthenia are not uncommon. In addition to the genital atrophy the following features have been mentioned as evidence of endocrine disturbances: relative lymphocytosis, eosinophilia, increased coagulation time of the blood, increase or diminution in the size of the thyroid gland and increased irritability of the facial nerve (Chvostek's sign).

A constant and characteristic feature to which the disease owes its name, is the myotonia. This may remain more or less generalized, but it may also be limited to only a few muscles. It is characterized by a slow relaxation of the muscle at the end of contraction; the phenomenon is best and most frequently shown in relaxing the hand-grasp. By persistent percussion of the involved muscles a mechanical myotonia is produced. This is most commonly elicited in the tongue and in the muscles of the thenar and hypothenar eminences, although it may be present in many other muscles.

Absence of demonstrable weakness in the orbicularis oculi seems to be the rarest negative finding, while absence of weakness in the sterno-

ceido-mastoids is next in the order of rarity. The extensors of the neck are weak, and therefore the head inclines forward, so that the eyes are directed towards the ground. This condition, associated with a drop-foot and steppage gait due to weakness of the muscles of the leg, presents a picture resembling the attitude and gait in tabes.

Speech is nearly always affected. It is low and monotonous, and has a more or less nasal quality. This may be due either to muscular atrophy, or to myotonia, or to both, for the lips, tongue, soft palate and vocal cords are often weak, and the tongue and the other muscles are frequently myotonic.

PARAMYOTONIA CONGENITA.—Eulenberg,²¹ Delprat,²² v. Sölder,²³ Senator,²⁴ Martius and Hansemann,²⁵ and others have described cases of myotonia in which a tardy relaxation of the contracted muscles occurred only in the cold. Eulenberg designated this special type of myotonia as “paramyotonia congenita.” It is a heredo-familial disease in which the

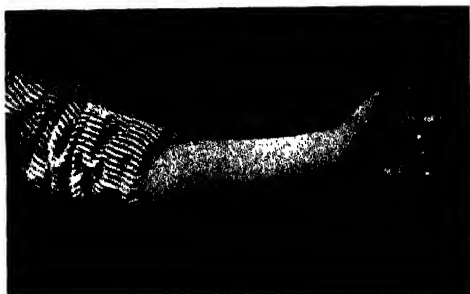


ILLUSTRATION 30.—PALMAR ASPECT OF HAND IN SISTER, SHOWING TYPICAL MYOTONIC REACTION (FIST ALMOST COMPLETELY OPENED). Note Fingers are Still in Flexion Though not as Much as in Illustration 28. (J. Neurol. and Psychopathology.)

myotonia seems to be confined to certain voluntary muscles of the face, especially the orbicularis palpebrarum and orbicularis oris, so that the patients are unable to open their eyes or to speak for a quarter of an hour or longer. The myotonia in these cases does not seem to be influenced by excessive exertion; cold brings on the spasm, and warmth has a tendency to diminish it. The muscles of the neck and of deglutition, as well as of the limbs may occasionally be involved.

The mechanical irritability of the muscles is not increased, but electrical examination shows a diminished excitability. The pathologic findings are similar to those of myotonia congenita. The characteristic symptoms of the disease make their appearance at birth. It has been found coincidently with Thomsen's disease in members of the same family.

Treatment of the Myotonias in General.—Gymnastics and massage have been employed by Oppenheim and Beehterew. Frink reports the administration of **thymus extract** in one case with good results. Gessler proposed, in cases with severe spasms, to bring about an atrophy of the muscles by **stretching the nerves**; his proposal has been rejected. Johnson and Marshall suggest the use of **strychnin**. In several of the

atrophic cases under the writer's care at the Montefiore Hospital, all forms of treatment—hydrotherapeutic, gymnastic, electrical, gland extracts, strychnin, etc.—have been employed without the slightest sign of improvement.

Prognosis of the Myotonias in General.—The prognosis as to life is good. The disease is chronic and progressive; remissions are not uncommon. The cases which have been reported as improved were undoubtedly atypical forms following trauma or infections.

Pathology and Pathogenesis.—The symptoms of myotonia have been reproduced in animals after poisoning them with veratrin and creatinin. Many investigators believe the condition to be due to an exaggerated excitability of the sarcoplasm. Erb and others have found an enormous hypertrophy of all the muscle fibers; in some cases these were twice the normal size. They also found a profuse proliferation of the nuclei in the sarcolemma, indistinct striation, vacuolization and a slight increase in the connective tissue. Some investigators found atrophic as well as hypertrophic fibers. Schiefferdecker²⁰ believes that the increase of the nuclei is only a relative one, but he was able to demonstrate in the sarcoplasm granules which are not seen in normal muscle tissue. Jacoby⁴ also found changes in the muscles, but he does not attach much importance to them from a pathogenetic point of view. Changes have also been reported in the central and peripheral nervous system, but not with sufficient constancy to be considered other than accidental or secondary in nature.

Various theories have been proposed to explain the pathogenesis of the disease: (1) Theory of cerebrospinal genesis (Thomsen, Engel, Curschmann, Johnson-Marshall²¹); (2) myopathic theory (Strümpell, Nissen¹¹); (3) theory of combined cerebrospinal and myopathic genesis (Findlay²⁸); (4) chemico-toxic theory (Bechterew, Jacoby⁴); (5) theory of endocrine dysfunction. A critical review of all these theories makes it obvious that the pathogenesis of this disease is still to be determined. By far the most pronounced feature of the condition is its hereditary character. The symptoms and course would seem to suggest as the basis of it some form of degeneration—a form of "*abiotrophy*." As a matter of fact, in spite of the more or less definite pathologic changes found in the central nervous system in the cases of so-called symptomatic myotonia, the mechanism of the phenomenon of myotonia is not at all clear. It is difficult to explain why a lesion in the lenticular nucleus or in the cerebral peduncle will give rise to myotonia in one patient while an identical lesion in another patient will produce no myotonia.

Historical Summary.—The disease was first described by Charles Bell as early as 1832, but was actually "discovered" in 1876 by the Silesian physician, Thomsen, who was afflicted with the disease himself, and in whose family he could trace it to 20 cases in five generations. Nissen, a grand nephew of Thomsen, could trace the disease in his own family through seven generations, in which up to 1923, 219 persons were affected. To Erb,²⁹ however, belongs the credit of having classically worked up the disease, especially as regards the electrical changes in it.

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MYATONIA CONGENITA (OF OPPENHEIM)*

History and definition, p. 580—Occurrence, p. 580—Symptomatology, p. 580—Diagnosis, p. 581—Treatment, p. 581—Course and prognosis, p. 581—Pathology and pathogenesis, p. 581—Bibliography, p. 583.

Synonym.—Amyotonia congenita.

History and Definition.—In 1900, Oppenheim drew attention to a condition characterized by marked muscular atony and a peculiar paralysis occurring in early childhood, and closely resembling the clinical picture of poliomyelitis.

Occurrence.—The disease is noticeable at birth, but typical cases have been known to have begun in the first and second years of life.

Symptomatology.—The involvement is most marked in the muscles of the lower, sometimes also of the upper extremities, and less often in those of the neck and trunk. Involvement of the head, face, tongue, eyes and larynx has not been observed. The muscles do not appear atrophied, but their flaccidity is very striking. The atony is so marked that the extremities can be placed in the most bizarre positions, as if they were loosely-attached appendages. When the little patient is seated the trunk bends forward, forming a marked kyphosis. That the paralysis is not a true one, but rather a weakness, can be demonstrated by placing the child on his feet, when it will be seen that not only is he

* This is not to be confused with myotonia congenita (Thomsen's disease), or with dystonia musculorum deformans, the three diseases being distinct conditions.

unable to stand alone, but even when supported, his legs give way under him; but when he is placed on his back, feeble active movements of the arms and legs can be carried out. The hypotonia of the various joints is also a very striking symptom.

The cranial nerves, the sphincters and sensation are not involved; there are no fibrillary twitchings. The deep reflexes are absent, while the superficial ones remain normal. The direct and indirect electrical excitability, both to faradism and galvanism in the paralyzed as well as the apparently sound muscles is usually reduced and frequently absent; there are no polar changes. In one of Oppenheim's cases, while the legs lay as if paralyzed, previously absent knee jerks could, after electrical stimulation, be freely brought out.

Purser¹ met a case of this disease in a two-year-old child, in which the condition developed at fifteen months, after an attack of severe diarrhea lasting four weeks. The patient had rickets and an unusual symptom—nystagmus. According to Purser, this was the first case of myatonia reported up to 1914 in Ireland.

Diagnosis.—(Collier and Gordon Holmes (cited below) point out the following clinical features of the disease which distinguish it from the *myopathies*: (1) The absence of any familial tendency. (2) The disease is in the majority of cases congenital, and in a small number of cases it appears suddenly and is fully developed after certain acute diseases. (3) The local wasting and weakness of an individual muscle or of a group of muscles, so characteristic of all forms of myopathy, are not met with in myatonia. (4) Affection of the periphery of the limbs, and especially of the intrinsic hand muscles, which is the invariable rule in myatonia, is of the greatest rarity in any form of myopathy. (5) Myatonia never spreads to regions previously unaffected, slow spreading of the affection from muscle to muscle being characteristic of all forms of myopathy. (6) The deep reflexes are absent from the beginning, but may reappear after improvement in myatonia, while in the myopathies they are present at first and slowly diminish as the condition progresses, and finally are lost, never to be regained. (7) There is a tendency to improve in myatonia, and in some cases recovery may ensue; this is never the case in myopathy.

The absence of atrophies or sensory changes, the generalized symmetrical involvement, and the electrical changes which can be seen in most *all* the muscles distinguish myatonia from *anterior poliomyelitis*, and from the peripherally and spinally induced *intrapartum paralysis*.

Myatonia must also be distinguished from *syphilitic pseudoparalysis* as described by Vierordt, and *muscular weakness due to rickets*, as well as from *joint hypotonia due to involvement of the capsules and ligaments of the joints* (Finkelstein).

Treatment.—General hygienic and tonic measures are indicated. Careful medication with **strychnin, massage and electricity** are resorted to. **Orthopedic appliances** may be necessary. Powis and Raper found after the administration of **bile salts or dried ox bile** an increase in muscle strength.

Course and Prognosis.—The course of the disease is slow; the prognosis as to life is good. In some of the cases, in the course of time, a more or less complete recovery may occur.

Pathology and Pathogenesis.—Spiller,² in his first autopsy, found

changes in the muscles only, and none in the nervous system. Baudouin⁶ found nothing characteristic for pathologic inferences. In another case Spiller and Griffith⁴ found atrophy of the muscle fibers much more marked in some bundles than in others. The atrophied fibers retained their transverse striations and showed an excessive number of nuclei in the sarcolemma; some of the muscle fibers presented fatty degeneration. These were the changes found in the muscles of the forearm; the atrophy was most marked in the calf muscles and least in the muscles of the back. Accompanying these muscle changes was a small spinal cord with scanty and atrophied anterior horn cells; the anterior roots were smaller than ordinarily and stained very poorly. One posterior tibial nerve was examined and also found much degenerated. The brain in this case was unusually large. Skoog, Reyer and Helmholtz found similar changes in the muscles "*in vivo*," whereas Bing found the muscles to be perfectly normal.

Collier and Gordon Holmes⁵ in a study of 2 cases found most of the muscle fibers smaller, which they say might be attributed, on the one hand, to a lack of development, or, on the other hand, to atrophic processes affecting them; but they are more inclined to believe, on account of the extraordinary irregularity in the shape and size of the fibers, that the condition was due to an atrophy. The atrophy and decrease in the number of fibers, in their opinion, sufficiently explains the palsy, while the fatty infiltration and the increase of connective tissue cells conceals in part the general atrophy.

De Villers⁶ had 2 cases which confirmed Concetti's conclusions based on 68 cases with 16 autopsies, that the cause of the disease is an arrest of development or retardation during intra-uterine life of the anterior horns cells of the spinal cord, and that the lesions may invade the cerebrum, cerebellum, peripheral nerves and muscles, but that there is a tendency to progressive improvement.

Oppenheim believes the disease to be due to an arrest of development of the muscle fibers. Marburg held that the cases were forms of intra-uterine poliomyelitis. The symmetrical development of the defects, in all cases, the development of some of the cases after birth, and the fact that in a good many instances the anterior horn cells were not involved, and in those cases in which they were involved there was no evidence of inflammatory changes, speak against the correctness of Marburg's theory.

Some investigators are inclined to ascribe the disease to a disturbance of the ductless glands. Berti considers it a variety of congenital myxedema. In this connection it may be of interest to note that Powis and Raper⁷ were led to the following conclusions from their metabolic studies in this disease: (1) There is a diminution in hepatic function, as evidenced by the presence of acholia; (2) normal calcium retention is associated with a relatively high potassium retention; (3) there is a low creatinin excretion with a relatively high creatin excretion.

Rothman believes the cases to be a type of Werdnig-Hoffman's disease; the fact that there is no wasting of the muscles and that there is a tendency to improve after a time, speaks against this contention. A. Gordon⁸ believes that there is an antenatal disturbance of nutrition of the anterior horn cells, which in some cases leads to their destruction, thus producing a Werdnig-Hoffman type; in other cases the nutritional

disorder is slight, resulting in a diminution of muscle tonus. He also believes that there are a large number of intermediary cases difficult to classify clinically.

Bernhardt is inclined to assume an injury to the peripheral nerves, somewhat like a generalized polyneuritis of autotoxic or infectious origin.

From the facts adduced it is evident that the pathology of the disease is not definitely known, and the same may be said of its causation. There is a tendency to consider the condition a type of myopathy rather than a distinct clinical entity.

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Diagnosis.—The disease must not be confused with organic nervous disease, such as tabes, disseminated sclerosis, hemiplegia, paralysis agitans and other conditions in which difficulty or inability to carry out complex movements is a prominent symptom. Writer's cramp has been wrongly diagnosed when the patient was suffering from agraphia, due to a tumor of the brain. A careful neurological examination with the finding of paralyses, sensory disturbances, electrical changes in the muscles or nerves, will make the diagnosis clear. The differentiation may at times be difficult when a hysterical or neurasthenic patient is, on account of tremor, unable to write.

Treatment.—The treatment to be carried out is with a view of improving the patient's general condition by **proper nourishment, hydrotherapy, change of environment** and other similar measures. Drugs seem to have little or no effect. Whenever possible, **absolute avoidance of the occupation** which brings on the neurosis is advisable. If for economic reasons this cannot be carried out, the patient is to be instructed to assume a **correct posture** while at work, and he is to be provided with **properly constructed tools or instruments**. **Massage, gymnastic exercises, and Swedish movements** are indicated. In writer's cramp, special penholders have been constructed which are held between the index and middle fingers (Zabludowski); a Nussbaum bracelet which makes the flexors and adductors superfluous during writing may be employed with benefit; in some cases the use of the typewriter may be necessary.

Course and Prognosis.—The neurosis may last throughout the patient's life. Some patients learn to write with the left hand, but sooner or later this hand also becomes affected. Recovery may take place, but relapses are very common. Like in all the neuroses, the longer the duration the worse the outlook for recovery.

Pathology.—No lesions have been found to explain the malady. The disease is considered by many to be a variety of exhaustion neurosis.

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CHAPTER XXII

THE TRAUMATIC NEUROSES

By EDWARD E. MAYER, A.M., M.D.

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This title is employed to designate any combination of nervous symptoms which follows an injury and which is not the result of organic lesions of the brain, spinal cord or peripheral nerves. The accident may not have produced any structural disability, yet the psychic trauma attached to it brings on functional symptoms. Generally, and mostly hysteric in type, partially neurasthenic and hypochondriac, occasionally accompanied by definitely psychotic phases, these symptoms were believed to marshal themselves in such a grouping that the traumatic neuroses were, and still are, accepted as a distinct entity. It was so regarded by H. Oppenheim,²⁴ who is responsible for the name. Many recent writers miss the fact, however, that he described *the* traumatic neuroses and not *a* traumatic neurosis. Grouped under this name are syndromes whose clinical signs do not conform to the classical pictures of hysteria and neurasthenia. In their elucidation we deal occasionally with the problems of a reflex epilepsy, of localized muscle spasms, and of tics of varied types. Even dystonia and myoclonia may enter into the group picture. Overshadowing the psychic symptoms loom frequently the problems of a psychopathic inferior make-up. And on the physical side, the question of actual concussion injury to the brain and spinal cord constantly presents itself. In fact, many authors believe that there is always some molecular or other form of alteration to nervous tissue, and that it is merely our ignorance to which is due our inability to find these changes. On the other side are those who believe in a psychogenetic origin to most of the traumatic neuroses.

It was an English physician, Erichsen,¹⁴ who in 1866 drew attention in a systematic way to the nervous symptoms found without any demonstrable causative injuries in victims of railroad accidents. He thought they were the result of concussion of the spine, and his so-called "railway spine" was believed by him to be due to minute, material alterations in the spinal cord. This opinion became so popular that it lived for many years after many observers, and especially Oppenheim in Germany, Page²⁶ in England, Walton and Putnam in America, and Charcot⁷ in France, definitely established the fact that most of the symptoms included under "railway spine" or spinal irritation were those of a psychoneurosis. Page went to the other extreme. He not only attacked Erichsen's views concerning an organic basis for the symptoms included under "railway spine," but also asserted that concussion without demonstrable injury never injured the spinal cord, except in the very rare symptom-complex of intraspinal hemorrhage. As Dana summed it up: "Concussion is mental shock and physical bruising." A little later than Page, in 1889, Oppenheim²⁴ published his well-known book, in which he insisted upon the occurrence of symptoms following injuries which differed from traumatic hysteria and traumatic neurasthenia. To this day we find two camps. One explains the differences in situational factors, influenced through suggestion; it adopts a psychogenetic etiology. The other is inclined to agree with Oppenheim; it postulates undetermined cerebral changes in at least a small proportion of the patients. The followers of the latter belief are, however, in the minority.

In the great war just closed, shell-shock, war-shock, and war-strain were words coined to describe the same group of symptoms which in civil life were included under the traumatic neuroses. The history of the traumatic neuroses repeated itself. These terms at first were thought to delimit a new type of organic disease. This idea did not, however, live long. Shell-shock is the traumatic neuroses of war. Environmental factors before, as well as determining psychic influences after the shock, tended naturally to introduce minor differences into the picture.

This incondite grouping—the traumatic neuroses—claims our attention, therefore, as a varied set of symptoms with some trauma in the background as the determining factor in the onset. Association with the accident implies a psychic shock as the origin, but not necessarily any injury to nervous tissue or even a physical injury. This is the background. In the foreground often looms the question with civilians of compensation, litigation and suggestion, and with soldiers of trench-fear and war-dread. In both classes malingering and simulation must be dealt with. A blanket-title like the traumatic neuroses denotes, therefore, to the writer all symptom-complexes referable to the nervous system not dependent upon organic lesions which have an injury or shock as the predominant, causal factor.

Etiology.—We are describing the neuroses resulting from trauma, and, therefore, it need hardly be said that an accident is the CAUSATIVE FACTOR. The individual may have been in an accident, however, without having any physical injury. The traumatic factors, in other words,

may be entirely psychic, consisting in the fear and fright of the accident, or in the belief that an injury has been entailed. Some individuals who are neuropathic and who have as a consequence a state of anxious expectancy, dreading the day when they will be in an accident, succumb instantly to the psychic situations surrounding it, and do not readily react from them. Such anticipatory neuroses were extremely common among the soldiers during the recent war. They need not have been predisposed by heredity. The intense strain they were subjected to, the exhaustion resulting from their duties, combined with the wish to be out of the firing zone or the constant fear of being injured, were sufficient reasons for the production of a breakdown of the nervous organization of the soldier.³⁰ Early in the war, some observers thought they were seeing new types of disease; but this opinion was soon disproved and the war-neuroses were found to be similar to the traumatic neuroses of civil life, colored differently by the environmental factors of warfare. Eder¹³ found that 30 per cent. of the war neuroses had an antebellum history of a psychoneurosis. Forsyth's and Wolfsohn's figures were even larger.

The increased industrial development of the age has produced innumerable kinds of accidents, any of which may result in the production of the traumatic neuroses. The development of compensation laws has called attention to their frequency. This has resulted in an increase in at least one kind of the traumatic neuroses, i.e., the expectation, or litigation-neurosis. All kinds of the traumatic neuroses develop after railroad and street-car accidents. Electric shocks from dynamos and defective wiring are also fairly common causes. These present more often evidence of organic involvement than do accidents of other kinds where a neurosis is diagnosed. The neuroses which follow operative measures, as, for instance, the removal of hemorrhoids, are regarded by some writers as coming under the category of the traumatic neuroses. Scar-tissue from either wounds or operations may bring in their wake sufficient irritation to cause a local neurosis or a reflex epilepsy of a traumatic nature. That thunderstorms or lightning may produce the traumatic neuroses is well known, either physical or psychic factors, or both, being responsible.

PREDISPOSING FACTORS in the etiology are the previous lowered resistance of the individual due to alcoholism, saturnism, arteriosclerosis, syphilis, night-working and poverty. Loss of sleep, poor food, wet quarters, the intensity of the life, were accessory factors in producing the traumatic neuroses of the war. It is a moot question whether occupational factors may be considered as traumatic. The risks of the occupation must be taken into account in deciding compensation whenever a traumatic neurosis is the diagnosis, independently of the consideration of poisoning from phosphorus, arsenic or lead, or dampness, as being etiological factors. They existed as a necessary part of the occupational hazard, and coupled with the occurrence of an accident, render the prognosis worse.^{10, 24}

Symptomatology.—Signs of disordered action of the nervous system, functional in type, may occur immediately after an accident or shock. This is especially true after head injuries. A previously unrecognized nervous liability of the individual, or an inadequate glandular efficiency brought into symptomatic expression by the emotional response to an unusual situation, are often responsible.¹⁸ Generally, however, there is a period of weeks or longer after the accident before the symptoms are noticed. Contemplation of the physical effects of the accident, and anxiety concerning its possible consequences, produce an ideational bias which results disastrously to the nervous system. Often the anticipation of injury from working among dangerous machinery, or as in war from being constantly in imminent danger, serves to create a situation which needs merely the actual occurrence of an accident to bring out a latent neurosis.

The symptoms of the traumatic neuroses must be dealt with, therefore, from various points of view. It will simplify matters first, to outline the more common symptoms which include those found in hysteria and neurasthenia; later, to describe the special syndromes which are considered by different writers as also falling into the category of the traumatic neuroses.

A. THE HYSTERICAL SYMPTOMS.—Reviewing, therefore, briefly the symptoms of hysteria, whether they are traumatic in origin or otherwise, we find that they include motor and sensory symptoms and disorders of the special senses, psychic symptoms, and vasomotor and trophic alterations.

(a) *Motor Hysterical Symptoms.*—These are both *paralytic* and *irritative*. Of the latter, tremors are the most common. There is, however, no distinctive hysterical tremor unless we wish to consider all emotional tremblings as hysterical. A dry cough, hiccoughs, *aërophagia* (air-swallowing), meteorism, abdominal spasms with false signs of a peritonitis, a torticollis or a trismus—all of psychic origin—are frequently observed. But those types of motor symptoms are not as common as are the motor paralyses and contractures of legs and arms.

The *motor paralyses* and *contractures* which occur in the traumatic neuroses are, as a rule, those of hysteria. They may involve one or both sides, but generally take in both lower limbs or one arm. A hemiplegia is, however, not uncommon. Unlike the organic kinds, it does not involve the facial or hypoglossal nerves as a rule, and is generally upon the same side as the injury. Likewise, in monoplegia, the left arm is much oftener affected than the right arm. The paralysis is generally a flaccid one, and may or may not be accompanied by contractures. The apparent broadening of the lower limbs due to outward rotation (Heilbronner's sign) is not found in a paralysis of psychic origin. The "nursing arm" type of hysteric brachial monoplegia, the dragging of the affected leg after the sound limb, without tilting of the body and with a stiff back in an hysterical paralysis of the leg, flaccid fingers with elbow and wrist contractures, the sudden onset of a contracture, movements of a paralyzed limb under anesthesia or alcoholism, and disappearance

of recently-established contractures during anesthesia, are finger-posts which reveal a functional type of paralysis. We also frequently find a complete flexion of a leg, the heel touching the buttocks; and in the arm, the strongly-flexed hand touching the sternum with the elbow rigidly adherent to the side of the abdomen.

Paralysis of a functional type reveals, in its very nature, its psychic origin in that it does not involve isolated muscles but coördinating sets of muscles which subserve volitional impulses. The muscle-masses generally retain their normal volume and are never the seat of degenerative atrophy. An extreme flaccidity is commonly found with undue contraction of antagonistic muscles, or contractions are noticed in an entirely different set of muscles upon apparent effort to use the affected muscles. Some lessening in the volume of muscle masses after long disuse may occur. Occasionally we find even an increase in volume. Apparently there is, however, a non-hysterical functional type of paralysis which presents both atrophic and vasomotor alterations. Babinski and Froment have described this under the name of *reflexe physiopathique*. We will discuss it later.

The lost or impaired use of a limb after recovery from the actual injury is frequently noted in the traumatic neuroses. A careful examination by the diagnostician, aided by a roentgenogram, will tend to establish the functional or simulating nature of such disabilities. The paralyzed limb is, as a rule, more inert and helpless than it is in the case of organic paralysis. Vigorous movements, however, bring out contractions; sudden excitement or fear often reveals the fact that movements are possible. Vasomotor symptoms are not necessarily a part of the picture in functional paralysis. Manipulation of an affected limb often reveals the functional character of the paralysis in that a momentary retention of a limb occurs in a position only possible through the use of the paralyzed muscle masses, or there occurs an overpronounced resistance to movement by contraction of the antagonistic muscles. This contraction is an important diagnostic point in differentiation both from organic disease and from malingering. The organic type of reflex changes do not occur in functional paralysis. An occasional writer has claimed to have observed a true clonus or a Babinski toe-reflex in hysterical types of paralysis, but such observations are not free from suspicion. The glossy skin of organic paralysis does not occur in hysterical paraplegia.

Contractures of a non-organic type are frequently found. They are not necessarily hysterical in nature. They may result after actual muscle injury and persist by stabilization of the affected muscles (nature's splint) even after the injury has disappeared. A psychic element akin to those of habit-formation of all kinds serves to fix the muscles. But such functional types are not as frequent as are those of a hysterical nature. That tubercular bone and joint disease often results from injuries and commence with defense contractures must not be forgotten.

An hysterical scoliosis is not uncommon. The scoliosis is unaccompanied by torsion of the vertebral bodies and often disappears when the

patient lies upon the abdomen. Also the curvature is always a single one, is generally located in the dorsolumbar region, disappears during sleep, and has come on suddenly without sufficient organic signs. An abnormal position of the scapula (simulating "angel-wing" paralysis) is sometimes found. A lordosis of an hysterical type is not common.

In all contractures of limbs after trauma it is important to insist upon a careful examination of the joints. Roentgenograms should always be made. The excessive pains radiating from the joint with their disappearance when the patient's attention is distracted, the character of the muscle spasms accompanying the outbursts of pain extending often over the entire limb, the absence of cellulitis or local elevation of temperature, the presence of hysterical attitudes, functional types of sensory alteration and other evidences of hysteria, should suffice to exclude true joint-disease. If continual observation is possible, relaxation during sleep is noted; manipulation of the limb will often not awaken the sleeper, which could not be the case if pain is evoked. Oppenheim, it is true, claims that hysterical contractures occasionally persist during sleep. If of long standing, so that myogenic changes have occurred, this is of course possible, but never in contractures of comparatively recent onset. Organic contractures of cerebral origin may persist in sleep, but these would hardly need to be differentiated from mono-articular disease accompanied by contracture. The resistance to movement in hysterical paralysis always shows to the trained observer the mental effort used in resisting the use of a limb.

A not uncommon type of functional motor paralysis is a disassociation of motor function, in which there is no impairment of movement of the limbs while resting in bed, although an inability to stand or walk is claimed. This abasia may be a partial one, called therefore a dysbasia. It presents, as a rule, incongruous features which reveal its psychic basis, often of a frankly expressed dread or fear. An over-emphasis in movements is noted in all attempts to walk or stand, with attitudes which betray psychic resistance. The toes contract and seem to stick to the floor, the patient holds his back and head more than erect, or he flexes his body to an extreme degree upon his hips. Another common type is accompanied by spasm of the muscles with gross tremors upon attempts to use the limbs. Extreme types of this were presented in the so-called dancing gaits which were common among the soldiers.

(b) *Sensory Hysterical Symptoms*.—Pain is an almost invariable symptom. The recession of the physical injury does not give the patient relief, but is succeeded by complaints of pain localized around the injured region or the presupposed injured region, or there are complaints of pains everywhere, exacerbating when the patient is under observation or reciting his story, receding when his attention is distracted—all colored greatly by the mood of the injured person. A piercing pain in the temples (clavus), a pressing pain over the vertex of the skull, coccygeal back-pains, intercostal pains, pseudo-angina pectoris, are the most frequent complaints. A hyperesthesia of the skin covering the cranium is an almost invariable symptom. Hemierania is often of hys-

terical origin, as can also be a neuralgia of the fifth nerve. Workmen seem to complain of back pains, no matter what the nature of the accident, if they become victims of a traumatic neurosis. And neuralgia of a visceral location also demands the frequent attention of the physician in the traumatic neuroses. It is needless to state how painstaking must be our efforts to eliminate all possible organic causes, and to weigh secondarily to these our positive signs of an hysterical origin. The occurrence of the pain only at night, disappearance upon distraction of the patient's attention and unphysiological innervations of the pains are not of much value in establishing a diagnosis, unless accompanied by other hysterical symptoms elsewhere, and positive elimination of local disease. Abdominal operations are frequently undertaken in response to the subjective complaints of hysterical patients. This is positive elimination. The writer favors such measures in doubtful types rather than that some organic disease just beginning should remain undiscovered. For material disease processes are often accompanied by hysterical phenomena.

The sensory anomalies of the traumatic neuroses are generally more extensive than subjective pain complaints or localized hyperesthesia. Loss of the sensation to touch and pain is rarely, however, of a complete type. Complaints of severe pain, accompanied by objective anesthesia and analgesia, more or less complete, is the characteristic type. This is the functional form of dissociated sensibility. It never conforms to the areas innervated by spinal segments or peripheral nerves. It is a hemianesthesia which ends sharply at the middle line of the body and involves the mucous membranes, joints, even the bones. Or it takes the form of a helmet over the head or a gauntlet of the arm or a stocking upon the leg. We often find intact tactile sensation accompanied by analgesia. The analgesia is rarely constant, altering its innervation at different examinations, especially so if there is a change in the emotional factors influencing the patient. It may spread over the entire body and it is usually accompanied by disorders of the special senses.

(c) *The Special Senses.*—Sight and hearing may be lost or decreased; smell and taste may be decreased or absent. The deafness complained of is generally bilateral. More often a hyperesthesia of hearing, as well as of smell and taste, is found. These alterations are generally upon the same side as is the hemianesthesia or analgesia, though they may be bilateral.

We do not find an actual loss of central visual acuity in hysteria. It is more often a concentric contraction of the visual fields with an inversion of the color fields upon perimetric examinations. Occasionally a total achromatopsia is found. In comparison with other hysterical disabilities, visual symptoms are, however, rare. A hysterical blind eye generally sees and can be made to acknowledge seeing something by testing it. An unusually mobile reaction to light and accommodation (hippus) is characteristic. The sclera and cornea take part in the sensory loss. Amaurosis and amblyopia of hysterical origin is, of course, never accompanied by organic lesions of the fundus. A careful refraction is necessary and is too often neglected in hysteria accompanied by am-

blyopia. A morning ptosis (Gowers) is not infrequently noted. This consists in an inability to open the eyelids upon awakening without resorting first to manual aid. Perhaps the most common symptom of the ocular apparatus is that of blepharospasm. The keynote of diagnosis is the detection of inconsistencies, not compatible with organic involvement.

A perverted sense of smell or taste is not unusually complained of in the traumatic neuroses. A hysteric type of deafness is not as common as is an hyperesthesia of hearing consisting largely of subjective noises and accompanied by anesthesia or hyperesthesia of the external auditory canal. Bilateral deafness rarely exists unaccompanied by mutism. In the functional type of unilateral deafness, which is rare, the patient does not incline his head to the other side to catch sounds and the use of Rinne's and Weber's tests (or of the Galton whistle and the monochord) reveals its true character.

A sudden total loss of voice-production, more often a loss of speech, except for whispering, is a most common type of hysterical paralysis. A careful laryngoscopic examination is necessary. Absolute mutism is rare. The hysteric nature of some forms of stuttering is well recognized. If present before an accident, it is made worse by it. [Such an exacerbation was offered in one of my cases as the basis for a claim for large monetary damages.] In all speech disturbances of hysterical origin, the over-use of gestures and grimaces is observable; more effort than in organic types is attempted in order to bring out words. The rapid succession of mutism after stuttering or *vice versa* (especially in recovery) is characteristic. A hysterical paraphasia sometimes occurs. [The author has recently observed this symptom in a patient who cut himself in an attempt at suicide after criminal charges had been made against him.] Hysterical types of bulbar paralysis (swallowing-spasms, dysphagia, phagophobia, etc.) are very exceptional symptoms.

Bladder paralysis with incontinence of urine has been reported as of hysterical origin, but it should be considered at least as very rare. Hysterical anuria is occasionally noted. The writer has placed patients with this symptom under most careful observation and has satisfied himself of its occurrence. An anesthesia of the mucous membranes of the genito-urinary organs invariably accompanies it. Pollakiuria or an intense desire for urination with frequent evacuation of small amounts of urine is common. A careful inquiry into all local sources of irritation is, of course, always necessary in such types of hysteric symptoms. Impotence is frequently complained of after injuries. A polyuria with glycosuria is often found in individuals who have other symptoms of a traumatic neurosis. The above symptoms, largely the results of emotional shock upon the vegetative nervous system, in fact were recognized long before the traumatic neuroses were established as a syndrome. Today, however, we prefer to separate them from true hysterical (suggestive) symptoms and include them under disorders of the vegetative nervous system.

(d) *Psychic Symptoms*.—All victims of the traumatic neuroses show the irritability and depression which characterize hysteria. Many of them have a loss of recollection for the immediate period following the accident (retrograde amnesia). But the characteristic depression of the traumatic neuroses, as well as the type of memory change, as a rule is somewhat different from other forms of hysteria. The dispositional depression is deeper. It is almost that of melancholia. It is accompanied by constantly uttered ideas of hopelessness or of invectives against those whom they regard as responsible for their injury. There may exist not only gaps in their memory, but memory-distortions also. They have forgotten any previously existing disease which they may have had; they have forgotten events which may have led up to the accident; they have lapses and fitful memories concerning recent periods also so that they cannot tell of occurrences since the accident. Throughout we generally can detect the over-emphasis, exaggeration, and the changeable moods characteristic of hysteria: The tendency to lies, the so-called *pseudologia phantastica*, is readily detectible. The depression which is almost invariably present is not of an hysterical type. It is rather the hypochondriasis of older writers, which we no longer regard, as they did, as a separate syndrome. Its definition by Gowers is “a morbid state of the nervous system in which there is mental depression due to *erroneous ideas of such bodily ailments* as might conceivably be present.” That part of his definition which is important to remember in connection with the traumatic neuroses has been placed in italics. It is often accompanied by an apathy and lethargy which are not hysterical, but abortive symptoms of a true manic-depressive psychosis. It is difficult, therefore, to judge whether such temperamental moods were present before the accident, or evoked by it. The lack of discriminating observation antedating the time of the psychic shock leaves a gap in our ability to analyze this factor with reference to time. Accompanying this depression, we often find somnambuloid or hypnoidal states. The patient will sit and day-dream with a drawn-haggard facies which betokens mental misery, and remain in fixed positions which sometimes are suggestive of the poses noticed in dementia præcox. An irrelevancy in answers to questions (*Ganser's symptom*) is occasionally noted. The development of a true psychosis in the traumatic neuroses preceded by, or accompanied by an hysterical symptom-complex, is not uncommon. The traumatic neuroses of the war just ended were frequently found to present this combination. Hallucinatory delirium of an hysterical type was a rather frequently observed symptom in the so-called “shell-shock” victims. This was to be expected because of the severe fear and fright factors accompanying their origin. Fear-states need not be ascribed to a neuropathy. They can affect any type of person. But the emotional upheaval must be intense, as in war, to break down the self-control of some. In the traumatic neuroses in general, however, it seems that the disorders of the autonomic nervous system which arise out of fear and fright occur rather disproportionately to the severity of the physical injury or the

apparent intensity of the fright. These symptoms are no more hysterical than they are neurasthenic.

B. GENERAL SYMPTOMS OF A NEUROSIS.—These include the various signs of altered functioning of the sympathetic and parasympathetic nervous systems. They are included in the older text-books among the symptoms of hysteria and neurasthenia. The influence which emotions have upon this part of the nervous system is, therefore, nothing new. More modern investigators following the lead of Hess and Eppinger have, however, enabled us to place the symptoms of the autonomic nervous system in a special grouping, and have broadened our viewpoint on the physiological side, especially with regard to the individual susceptibility to drugs as also to emotional states. The vagatonic syndrome includes especially bradycardia, bronchial asthma, hyperacidity, pylorospasm, cardiospasm, mucous colitis, spasms of the unstriated muscles of the gall-bladder, salivation, hyperidrosis, urticaria, etc. These symptoms often form a part of the status thymicolymphaticus, a syndrome which is often unrecognized in early life, or mis-called neurasthenia. Trauma is the frequent precipitating factor in bringing it into activity. Enuresis and lymphoid tonsils will frequently be noted as having occurred in early life and later the fatigability, muscle irritability, excitable reflexes and phobias of so-called neurasthenia, are present, accompanied by a low blood-pressure and a low blood-sugar content. The types which present this syndrome among the traumatic neuroses have not as a rule progressed to where we find pronounced infantilism, epileptiform spells, or marked skeletal defects. The problems connected with the relations between sympathetic mechanisms, endocrine functioning and emotional factors, are not easily separated from each other; in no group of individuals must their interdependence be more recognized than in the traumatic neuroses.

A large number of symptoms frequently enumerated under hysteria and neurasthenia are then referable to the autonomic nervous system. This consists of two divisions: (1) The cervicothoracolumbar sympathetic, and (2) the craniosacral sympathetic or parasympathetic. A physiological balance must be struck between the two to give us normal functioning of the heart, lungs, abdominal viscera, and genito-urinary organs. Over-activity of the craniosacral innervations gives us the syndrome of vagotonia. A lowered threshold to responses on the part of the sympathetic system proper evokes the syndrome of sympathicotonia. These syndromes have been studied in connection with the pharmacodynamic action of certain drugs; a partial similarity of results from over- or under-activity of various integral glands has been much discussed; the effect of the emotions upon these glandular secretions has been established. The individual often remains, however, when viewed only in the light of his physiology and anatomy, an enigma. We will discuss later the personal factors which influence these physiological reactive signs and symptoms. The symptoms (after Barker⁵) include:

(a) Ocular Signs.

1. Myosis.
2. Mydriasis.
3. Accommodation spasm.
4. Accommodation paralysis.
5. Widened and narrowed eyelid slits.
6. Von Graefe's sign.
7. Stellwag's sign.
8. Infrequent winking.
9. Exophthalmus.
10. Enophthalmus.
11. Epiphora.
12. Argyll Robertson pupil. (?)
13. Anisocoria.
14. Scotomata.
15. Positive adrenalin mydriasis.

(b) Respiratory Signs and Symptoms.

1. Laryngismus and laryngeal crises.
2. Asthmatic attacks.
3. Vasomotor coryza.
4. Pulsus irregularis respiratorius.
5. Oculocardiac reflex or Aschner's phenomenon (arrest of respiration in expiration with slowing of pulse, produced by pressure on the eyeballs).

(c) Circulatory Signs and Symptoms.

1. Tachycardia.
2. Bradycardia.
3. Pulsus irregularis trasystolicus.
4. Vasomotor angina.
5. Changes in blood-pressure.
6. Peripheral hyperemias and anemias.
7. Intermittent claudication.
8. Acrocyanosis.
9. Dyspragia intermittens intestinalis.
10. Changes in conduction time (dromotropic disturbances).

(d) Gastric and Intestinal Signs and Symptoms.

1. Excessive salivation and xerostomia or dry-mouth.
2. Esophagismus.
3. Cardiospasm.
4. Hyperacidity.
5. Achylia.
6. Gastrosuccorrhea.

7. Pylorospasm.
8. Gastrosplasm (anorexia nervosa).
9. Gastric atony.
10. Spastic constipation.
11. Nervous diarrhea.
12. Mucous colitis.
13. Meteorism.
14. Spasm of the rectal sphincter.

(e) *Symptoms and Signs in the Cutaneous System.*

1. Goose flesh.
2. Hyperidrosis.
3. Bromidrosis.
4. Pallor (vasoconstrictor).
5. Erythemia (vasodilatation).
6. Dermographismus (skin-writing).
7. Urticaria.

(f) *Symptoms and Signs in the Urogenital System.*

1. Anuria.
2. Urinary retention.
3. Incontinence (nocturnal enuresis).
4. Pollakiuria (frequent urination).
5. Tenesmus.
6. Renal colic (Dietl's crises).
7. Disorders of libido, of erection, of orgasm and of ejaculation (impotency, spermatorrhea, vaginismus).
8. Uterine atony.
9. Excessive menstruation.

(g) *Symptoms and Signs Referable to the Hemopoietic, Metabolic and Endocrine Organs.*

1. Status lymphaticus of Bartels with various growth and skeletal alterations, including
2. Increased or diminished glucose-tolerance.
3. Eosinopenia; also
4. Eosinophilia.
5. Lymphocytosis.
6. Steatorrhea.

Freud recognized the difference between purely psychic reactions and those of a more or less somatic basis, and grouped together the results of anxiety under the name of the *anxiety neurosis*. He placed under this title the general symptoms of irritability, anxious expectancy and situational fears, with disorders mostly of the autonomic nervous system, listing them as follows:

1. Disturbances of heart action, such as palpitation with transitory arrhythmia, and protracted tachycardia.
2. Disturbances of respiration—nervous dyspepsia, asthmoid attacks, etc.
3. Profuse perspiration.
4. Trembling and skin attacks.
5. Excessive appetite, often combined with dizziness.
6. Suddenly appearing diarrhea.
7. Dizziness on attempts at walking.
8. Vasomotor skin-reactions.
9. Paresthesia.
10. Nocturnal fright.

From these results of anxiety, there develops according to Freud various phobias, characterized in one group by fear of the environment, and in the other group by anxiety because of vertiginous attacks, or with vasomotor reactions because of the consequent fear of walking. The phobias of repression or substitution are not included here by him, but are grouped under the compulsion neuroses. These differ from hysteria, according to Freud,¹⁰ in that we have no conversion of the repressed emotion into physical symptoms; instead a displacement of the emotion takes place, attaching itself to other ideas in the mental life of the individual. This is, therefore, a defense neurosis. It corresponds in symptoms with the psychasthenia of Janet who classifies psychasthenia under:

1. Obsessions.
2. Pseudo-hallucinations.
3. Abnormal impulses.
4. Mental manias.
5. Ruminations.
6. Tics.
7. Forced agitations.
8. Phobias.
9. Deliria of contact.
10. Anxiety states.
11. Sense of strangeness and unreality.
12. Phenomena of depersonalization.

Phobias he groups into:

1. Algas and bodily fears.
2. The fears of contamination.
3. The fears of situation.
4. The fear of ideas.

C. THE NEURASTHENIC SYNDROME (*Exhaustion Syndrome*).—Despite the above-mentioned viewpoint, there remains to be considered an exhaustion neurosis which we can call neurasthenia and which often fol-

lows trauma, particularly head injuries. A conception of asthenia marked by fatigue, induced by night-work or even by living itself, and evidenced by symptoms of nervous instability, is still prevalent under the name of "neurasthenia." The keynote is an irritable weakness with responses of unrest, distaste, annoyance, anger and anxiety to very slight environmental factors. Heart, respiration and stomach give evidence also of this irritability. Headache, insomnia, vertigo, loss of weight and a vasomotor instability of the skin are almost constant symptoms. The ability for mental concentration is weakened; introspection becomes a fixed habit; all efforts at work are accompanied by great restlessness and distress of mind. Fixed ideas of serious illness are aggravated by the alarm experienced over the cardiac and abdominal symptoms which are present. In the traumatic forms, brooding over injury may lead to obsessions. Ergographic tracings reveal the irritability and proneness to early exhaustion; pulse records give evidence of arrhythmia and tachycardia; a fine rapid tremor is almost constantly noted; the tendon reflexes are exaggerated; muscle-tapping produces a quick and overactive response; an alimentary glycosuria is frequently present, or an oxaluria. We find also an increased perception to pain upon testing the faradocutaneous electrical reactions; and lastly, other vagotonic symptoms enumerated above may be present.

D. SPECIAL SYNDROMES AMONG TRAUMATIC NEUROSES.—(a) *Vasomotor Syndromes*.—Vasomotor symptoms were of frequent occurrence in hysteric and neurasthenic symptom-complexes of former writers. The right to group them in this way is questionable. Head-traumas, however, frequently give rise to a special combination of vasomotor symptoms which have been described as a concussion neurosis (commotion-neurosis). We are not referring to individuals with actual fracture of the cranial vault. Absence of definite organic symptoms places them in a functional category, although finer molecular changes are not positively to be excluded. Many autopsies have confirmed the opinion that they are really organic in origin. The trauma is generally accompanied by slight symptoms of concussion: vomiting and nausea, a temporary dazed state, perhaps a transient unconsciousness with a slowed pulse, a fragmentary amnesia, etc. Later there develops a "cerebral symptom-complex" (Horn¹⁹), consisting of head-pains with various kinds of perverted sensations around the scalp and in the head, vertigo, noticeable particularly upon stooping or in turning the head and accompanied by suffusion of the conjunctiva, congestion of the face, and a rapid heart. An intolerance to alcohol, a tendency to cry, or sudden outbursts of temper (Kaplan's explosive diathesis), accompanied by an impairment of the power to receive new ideas or to initiate any action, are noticed. Various ocular and auditory perversions and hyperesthesiæ are present also, especially attacks similar to Ménière's disease and to migraine. The use of Bárány's and other modern tests of the cerebello-auditory apparatus, lumbar punctures and carefully made skiagrams will undoubtedly establish an organic basis for many of this type of the traumatic neuroses. This syndrome is often spoken of as the vasomotor

symptom-complex of Friedman,¹⁷ who separated it from the psychogenic forms of the traumatic neuroses. It bears a close resemblance to the set of symptoms due to serous meningitis (Quincke). It has not been a rare thing in my experience to find this "neurosis" after head-trauma and the author is of the same opinion as Friedman and others that it is not a neurosis. The combination of head-pains, vertigo, alcoholic intolerance, irritability of the special senses, ear-labyrinthal symptoms with decreased ability to retain new ideas (Merkfähigkeit) and to comprehend new ideas (apperception) are the six primary symptoms of Friedman. After from one to three years, 60 per cent. recover and the others are grouped by him into four subdivisions:

1. The traumatic neurasthenia group.
2. The vasomotor symptom-complex group with constant headache and severe migraine attacks.
3. Those who present permanently delayed mentation and decreased intelligence.
4. A secondary dementia of a partial type producing more the picture of a feeble-minded child. This type usually presents a total amnesia immediately after the injury. Friedman bases the inclusion of this group upon only one case. The author has seen it several times with this modification, namely, that the apparent feeble-mindedness has not been any more permanent than have been the amnesiæ.

The recent war has produced many reports of this cerebral symptom-complex with much discussion concerning its organic or functional basis. The problems connected with the use of high explosives complicated the earlier surveys of such patients. Whenever, as is often the case, we find with the above symptoms, actual evidence of organic disease, as for instance a hyperemia of the optic nerve, a nystagmus or a transient aphasia, the diagnosis of the traumatic neuroses cannot, of course, be entertained.

(b) *The Emotional Syndrome*.—We find frequently individuals who are known not to have been injured but who express great fright by symptoms which may continue a long time after the accident or source of the fright has disappeared. Grasset has written a good description of the mimic phenomena of emotion in the war expressed by trembling, shrieking, attempts to flee, intense perspiration, etc. Similar pictures were found in the sufferers from earthquake shocks in different parts of the world. Dupré¹² has attempted a special classification under the name of "the emotional constitution," which is, according to him, a state of high emotivity due chiefly to trauma and resulting in deficiencies of functional equilibrium. The symptoms enumerated by him are principally those of disordered functioning of the autonomic nervous system, coupled with, on the psychic side, the various obsessions, phobias and psychosexual aberrations. It is hardly a distinct entity. He is grouping various defense reactions which prevent a breaking-down of the instinct of self-preservation into a conception of a disease process by joining them to somatic symptoms.

(c) *Reflex Neuroses of Traumatic Origin—Localized Neuroses.*—Many accidents occur with slight muscle-bruises, ligamentous and tendon wrenching, myositis or fibrositis. After all symptoms and signs of these conditions have disappeared, pain and immobility still persist. Or pain and immobility based upon an accident occur without any cause for these symptoms being discoverable. There is no class of patients more difficult to classify. Grouping the functional types under the localized neuroses we find the following syndromes:

1. *Akinesia Algera.*—Möbius described a limitation of movements due to pain under this name, because, not only no physical cause for the pain exists, but also other signs or symptoms reveal that it is a pain-hallucination, or *psychalgia*. It is, therefore, from the very nature of things a common enough symptom of the traumatic neuroses. When it is the only symptom it is difficult to decide whether it is simulated or not. [A girl slipped upon the car-step on getting off a car. Upon her complaint of pain, her knee and lower leg were bandaged by a physician. The pain persisted and the other leg was favored in movements for many months, although no physical injury could be discovered upon careful examination. The fear of being disabled and of not being able to earn a living, became added to the suggestion of real injury by the fact that the physician bandaged her limb. Later a third factor came into play through her attorney, who sued the street car company for large damages. For many months there existed an inability to use her leg properly, because of the pains produced by its use. Psychotherapy promptly effected a cure.]

A pain-hallucination of a similar connotation is often noticed in conjunction with one of the special senses. Pain upon using the eyes not due to muscle-imbalance or other local cause is a common symptom of all neuroses. Pain in the head from the ordinary sounds of conversation is also a not infrequent complaint. We may find present a desire to stay in bed because of the distress occasioned by the ordinary efforts of going around the house. This has been described as *atremia*. It does not differ in conception from the various place-phobias, and the writer finds it, as do others, often accompanying them in anxiety-states.

2. *Akinesia Amnestica.*—Under this title, attention was called by Oppenheim²⁵ to a flaccid, atonic paralysis, differing from hysterical types, not due to discoverable organic causes, favoring the left side, accompanied by trophic disorders of various kinds and resulting after trauma. Oppenheim attempted to build up a definite conception upon the physiological side of physical results of trauma by reason of overstimulation of cerebral centers. He reasoned from such overstimulation an inhibition to motor impulses which has no ideational or suggestion components, but which is altogether psychogenetic in origin. This is rather a vague and artificially created conception.

3. *Physiopathic Disorders of a Reflex Nature.*—Babinski and Froment² attempt to separate from among the traumatic neuroses a localized type which they claim is due to causes not psychic but physiological. They call them "*troubles physiopathiques d'ordre reflexe*," and

consider them due to trauma by reason of an over-stimulation of nerve-cells and tracts. They reason from their premise of over-stimulability an inhibition to motor impulses which has no suggestive or ideational components in its genesis. This is the *syndrome physiopathique*. Babin-ski,¹ who has many followers, contended many years ago that hysteric symptoms are due to suggestion, and, therefore, amenable to persuasion. As a result of his war experiences, he has set apart from these phenomena, which he styled "pithiatism," this syndrome which he and Froment call physiopathic or nervous disorders of a reflex type. They present no signs of organic involvement. In common with hysteria, the symptoms do not result from minor physical injury; do not correspond to known anatomical innervations; are difficult to cure; and are not amenable to counter-suggestion. In the complete syndrome are found muscular atrophy; increased tendon-reflexes; decreased skin reflexes; hypotonus; lowered mechanical excitability of the muscle masses with muscle flabbiness; decreased quantitative electrical reactions of the muscles, and occasionally of the nerves; hypothermia; various vasomotor disorders (cyanosis, a salmon-red discoloration, etc.); and diverse trophic disorders of the bones, ligaments, skin and nails. American as well as some French and English observers do not accept this division, but regard Babinski and Froment's type as similar in symptoms to hysteria, similar also in treatment and not incurable therefore. The dynamic conception with an attempt at a physiological understanding of the symptoms marks, however, a new viewpoint.

4. *The Akro-neuroses*.—There are many types of vasomotor and trophic disorders which follow trauma, and which cannot be grouped with the ordinary types of the psychoneuroses. We have just called attention to a recent conception of a group of these under the name of the physiopathic syndrome of Babinski. Akroparesthesia, or pain perversions, occur after trauma, as well as from other causative factors, and have been considered by some authorities as being due to minor neuritic changes, especially of the sensory fibers (Cassirer). Erythromelalgia may result from trauma but rarely gives rise to the diagnosis of a traumatic neurosis.

5. *Reflex Epilepsy*.—That epileptiform attacks may follow local injury, particularly if a callus or a foreign body is present, is well known. We refer to peripheral irritation and not to meningeal adhesions. Emotional states—fear and anxiety—generally accompany the traumatic causal factor and cast doubt upon the real nature of the attacks. A thymicolymphaticus syndrome should always be looked for in reflex-epilepsy.

6. *Localized Muscle-spasms—Tics*.—Although localized tics and muscle-spasms frequently follow trauma, they may be due to various local irritations or to psychogenic causes. A dynamic basis (nuclear irritation) is assumed by some. Brissaud and Meige have described the distinguishing signs of reflex spasms and separated from them the tics of psychic origin. With the latter, we must look for ideational factors expressed by defensive movements, the impulse to their excitation being

irresistible, and which if not carried out produces discomfort. No external stimulus is present and the movements are unlike those due to local causes (Barker⁵). They protect the patient from the effects of the accident until the memories of it are eradicated. Later this protective mechanism itself becomes fixed and must be broken down.

E. THE TRAUMATIC PSYCHOSES.—Though we are not considering psychoses in this article, it would be incomplete without some reference to them. The line between psychic and psychotic symptoms is not a sharp one. Hallucinatory deliria may occur in severe hysteria. A mild hallucinatory delirium with stupor on the other hand occurs often after head injuries, especially in elderly or in alcoholic patients. A memory defect for recent events as a rule is its termination and gives us the setting of a Korsakoff's psychosis. Generally a true psychosis eventuates long after the trauma, because personality (except upon the emotional and vasomotor side) does not alter suddenly and permanently, unless material injury to the brain tissues has taken place. In the latter, two types of symptoms are produced: those due to edema and inflammation, and those resulting later from scar tissue, sclerosis or degeneration. In the latter group belong posttraumatic epilepsy, multiple sclerosis and paralysis agitans. In the psychoses which slowly develop after trauma in individuals who presented hysteroid, epileptoid or neurasthenic symptoms earlier in the history, a careful study of the history generally reveals evidences of a psychopathic constitution existing before the accident. A dysthymia (moodiness), a zykllothymia (mild cycles of elation and depression), a querulent personality, are present long before a trauma has precipitated the onset of a true psychosis. An inadequate reaction to trauma with a resultant maladjustment is almost inevitable with varying phases according to the intensity and quality of the psychic factors involved. It is in this light that dementia præcox, manic-depressive insanity and paranoia following trauma must be viewed. Without a previously existing psychopathic constitution, the trauma in itself would not evoke these types of psychosis. There is, however, an acquired psychopathic constitution which follows trauma, regarded by some writers as the traumatic neurosis and is variously called situational-neurosis, compensation-neurosis, expectation-neurosis. Brissaud described it under the name of *sinistrosis*.

F. THE TRAUMATIC NEUROSES.—In this type of the traumatic neuroses, no doubt is possible of the absence of organic symptoms. Evidences of a functionally disordered nervous system—motor, sensory, or autonomic—are negligible. The entire picture is dominated by the mental attitude of the patient. He is entitled to damages and large damages; he nurses a conviction of the injustice done to him with the chance now of getting even. This idealogue is easily formulated, fixed and accepted in those who have had to work hard to barely support a family, who dread as much their inability to continue such support as any fear of illness itself, especially when friends suggest the opportunity to gain a monetary advantage from their misfortune.* Some emotional reaction must inevitably exist from a trauma. Insomnia, fright dreams, palpi-

tation of the heart, dermatography, and other physical evidences of this condition would terminate with the recession of memories of the accident, did not expectation of gain, and the worries of litigation, coupled with the suggestions of the physician and insinuations of the lawyer, serve to keep them alive. The writer prefers to separate these two factors resulting from the trauma, classing the immediate or directly emotional results as psychogenous, and the indirect results prolonging, accentuating, and specializing this type of neurosis as ideational. An innate susceptibility or predisposition need not be considered here as in the other types of the traumatic neuroses with hysteriform and neurasthenic syndromes. This type of neurosis is also called *litigation neurosis* or *compensation neurosis* by those who see only conscious deceit in its origin. Between intentional simulation and unintentional deceit no one can always draw a line. It is a social diagnosis as much as a medical one which is required.

G. CORRELATION AND VALENCY OF SYNDROMES.—In order to grasp the traumatic neuroses, it is necessary to classify them in the manner attempted above. Several points stand out clearly: (1) that whenever we have definite signs of organic disease, we should no longer classify the patient in this grouping; (2) that some patients are kept in this grouping who, had we more exact interpretations of some doubtfully organic symptoms, would probably be removed from it; (3) that we find in the traumatic neuroses a group whose only claim to medical attention is an obsession to recover damages and in whom symptoms of structural or physiological imbalance are wanting. There remains still the larger group of the traumatic neuroses whose symptoms are both physiological and psychological, i.e., who present psychic anomalies as in hysteria, and on the lower level of physiological innervation, evidence of defective activity of the autonomic nervous system.²⁰ The combination of so-called hysterical and neurasthenic syndromes cannot be understood, unless we attempt to view them in some fundamental way, in order to correlate the relations between the affective (emotional) traits of individuals and particular autonomic segments, whose function is to use sensorimotor stimuli in adjustments to environment, as well as to utilize the biologically established stimuli which have become conditioned, stabilized and automatic. We come to recognize in this way that a mere recording of the number of times dermatography, palpitation of the heart, tremor, analgesia, insomnia or paralysis are present in patients does not help. We must search for explanations of their occurrence and formulate causal relations based upon fundamental conceptions in order to estimate the valency of symptoms.³⁰ We get in this way to see why the traumatic neuroses do not differ essentially from other neuroses. In fact, their infrequency after accidents among employees of corporations bears witness to the insecure causal relation between injury and neurosis, as compared with their frequency among "injured outsiders." The statistics of Biss, covering almost 20,000 injured among employees of corporations, show a compensation list of 2,670, of whom only 12 or .45 per cent. were classified under the traumatic neuroses. Horn,¹⁹ in

1911, among 2,232 injured railroad employees, reports only 30 who were diagnosed traumatic neurosis. In the same year, in the same district, there were 195 injured in railroad accidents, of whom 89, or 46 per cent., were functional nervous disorders.

Pathological Anatomy.—The traumatic neuroses in the strictest sense do not include any disorders which are due to demonstrable pathologic alterations. The title itself indicates this. Autopsies as a rule confirm this. The findings of gross anatomical lesions speak for a wrong diagnosis *intra vitam*. Nevertheless, in the group of the vasomotor neuroses particularly, it is sometimes difficult to decide whether actual injury to cerebral, especially vascular structures, has occurred. Friedman regards his concussion neurosis as definitely due to disease of the arterioles. The neurosis symptoms in other groups may overshadow signs of organic injury which otherwise would be detectable and not ignored. Oppenheim, Vibert, Knapp, Crocq, Mott and others, it is true, regard the traumatic neuroses as having a pathologic basis; they premise our inability to detect the fine molecular alterations of the nervous system which have resulted from the trauma as being due to our present inefficient methods. Yet these observers are compelled to acknowledge the occurrence of the traumatic neuroses from fright without any actual physical trauma. We have mentioned before that Babinski, considering traumatic hysteria as entirely due to suggestion, sets up a claim of molecular changes in the group of the traumatic neuroses which, under the name of reflex physiopathies, he has separated from hysteria.

Schmaus, in 1890, described a necrosis of the axis-cylinders which occurred long after the trauma in spinal concussion, unaccompanied by any evidence of gross anatomical lesions before death. Bicheles, and more recently Allen, have shown that concussion produced experimentally upon animals may cause a degeneration of the nerve-fibers of the medulla oblongata and spinal cord. Evidence of organic lesions from alternating electric currents is offered by various observers. Many of the symptoms after severe electric burns indicate organic lesions of the nervous system. Slight electric injuries may, of course, result in a psychogenetic neurosis without physical or molecular changes in the nervous system. The dynamic conception of a psychogenetic syndrome, without there being, of necessity, any molecular changes in the cells, is a viewpoint which has met with much antagonism. A. Meyer,²³ an American leader in psychiatry, contends that we are on surer ground by viewing mental facts as such, without considering the possibility of "lesion" underlying them. The opinion of the opposite school has been expressed recently by Mott, who says "that although there may be no discoverable lesion in a 'psychic trauma,' yet so complex is the structure of the human central nervous system, and so subtle the chemical and physical changes underlying its functions, that because our gross methods of investigating dead material do not enable us to say that the living material is altered, yet admitting that every effect owns a cause, a refractory phase in systems or communities of functionally correlated neurons must imply a

physical or chemical change, and a break in the links of the chain of neurons which subserve a particular function."

Pathogenesis.—Oppenheim's conception of the traumatic neuroses is that they result from a combination of both physical and psychic shocks. "Both," he says, "act upon the cerebrum and produce molecular alterations in those areas which control the higher psychic spheres and those motor, sensorial, and sensory functions which are connected with them. A peripheral injury may likewise be transmitted along the sensory tracts and evoke the same alterations or an irritation arising from a scar may permanently influence the brain." That in the vasomotor neuroses we are dealing with structural alterations, especially of the arterioles which are responsible for many of the psychic symptoms we have already alluded to. Sollier, in his conception of *cénesthesia*, rears up on the sensory side a conception of the pathogenesis of hysteria, including the traumatic types which answers to the second part of Oppenheim's conclusions. A neuropathic disposition is accepted as existing, though often latent, and the effects of fright and other emotional factors are present in addition, and influence the character of the symptoms present. As recently as last year, Oppenheim has in a monograph again insisted upon this viewpoint. Lewandowsky's opinion that "a mechanical shock to the nervous system by way of the sensory and sensorial tracts is unknown to physiology and pathology" is accepted by most writers who believe that the traumatic neuroses represent a psychogenic syndrome only, and that all observations otherwise should be regarded as proof of an organic disease existing alone or in conjunction with a psychic disorder.

Wilson ventured the following explanation of the traumatic neuroses which he met with in the war: (1) at the synapses, there is a structural discontinuity of the nerve-fibers; (2) the results produced by impulses traveling in a nerve depend on the way the fiber ends, and not on any differences in the impulses themselves; (3) at the synapses different physiological symptoms come in touch with one another, and so co-ordinate action in diverse systems is possible; (4) at the synapses there is always a spread of nerve impulse and the greater the impulse the greater the spread. He concludes, therefore, that high explosives produce a dissolution of the permanent auditory pathway and a spread of nerve impulses into the other adjacent pathways. The auditory impulse no longer reaches its goal and deafness results. A similar dissolution may occur at any one or at all synapses. Functional inhibition, producing stoppage of function, and increased activity of function of other innervations are not satisfactorily explained, however, by such a hypothesis.

Monakow's conception of *diaschisis* offers a way of approach. In actual brain injury from hemorrhage and thrombosis, the primary symptoms are recognizably greater than are the symptoms due to the actual lesion. Surrounding areas are arrested in function for a time, to later take up again through various association tracts, their various functions. This inhibitory influence is spoken of as *diaschisis*. Shock without physi-

cal lesion may also effect a stoppage of function. The diaschisis theory of Monakow, therefore, need not apply only to organic injury. A stoppage of cerebral activity, not structural in origin, is well-recognized even by the layman who speaks of being "paralyzed from fright," "stricken dumb with fear," and the like. This dynamic influence of emotion is universally accepted. The writer is not ignoring the viewpoint of those who believe that the cortical effects of emotion are secondary to autonomic reactions, though he does not entirely accept it. The continuance of these results of psychic shock from either viewpoint depends upon the adjusting ability of the individual affected. Motor responses to emotional reactions will not subside if the situational influences are not removed. The idea of serious injury, accompanied or not by some physical trauma, is sufficient to prevent the return of a normal psychic life. The instinct of self-preservation is so affected that continuous emotional factors come into play and prevent adjustment to the otherwise minor results of the accident, producing the so-called defense reactions and protective phenomena. A predisposition to such influences as understood under the conception of a neuropathic constitution need not be present if the fright and fear effects are intense. The war-neuroses have shown this. The various hormone producing organs are stimulated to over-activity by the draining of neural energy from emotion. This over-stimulation of the glandular secreting organ produces similar non-integrations and symptoms such as those evoked by primary disorders of the endocrine organs. In their turn, these symptoms, interpreted as being the result of the accident, produce a vicious circle, marked by disorders of the autonomic nervous system on the one side, and by motor and sensory symptoms of a hysteric nature on the other. Neither the nature nor the severity of an accident predicates, therefore, the degree of emotional reaction. This, to recapitulate, is determined by the constitutional make-up of the individual, the integration of his ductless glands, the activities of his autonomic nervous system, and the innervations of his cortical centers. Actual vascular changes or chemical injury of a molecular nature may occur and influence the cortical cells and tracts, but without the factors of emotional disintegration of the glandular and autonomic systems they would not of themselves produce the traumatic neuroses. These cortical effects are transient, if the cells quickly reestablish their function; permanent, if the synaptic connections cannot become reestablished (Sherrington).

The writer feels that much of the confusion which exists concerning the cause of the traumatic neuroses is due to an over-emphasis of one or other of the above factors. In the above explanation the writer has purposely left aside various psychogenetic influences which cannot possibly be ignored in any complete survey. An analysis of every individual will bring us closer to an understanding of how the symptoms are produced. Not all emotion is frankly expressed by fear-reactions. The effort to suppress them, for instance, in the war just ended produced many a psychoneurosis, especially among the officers, though the common soldier was not unaffected. Emotion which does not have a direct sur-

face play brings out on the physical side substitutive symptoms and shirking reflexes. If of old standing, the trauma gives occasion to an intense explosion of these buried emotions which are converted into severe functional paralyses, both sensory and motor. Symbolism plays a strong rôle in normal life, but even stronger in these reactions precipitated by the trauma into full activity. The influence of submerged emotions varies immensely with different individuals, but it always has a dynamic effect upon their psychic life. The author is in agreement with Dearborn, that in all these processes the cerebral cortex is activated, since it represents the adjuster mechanism of the afferent neurokinesis. Pawlow's conditioned reflex studies on the physiological side help us to understand how these psychokinetic equivalents act, as Adler's organic inferiority conception on the psychological side offers evidence to the presence of psychic strain from childhood. His contribution concerning the defense neuroses is a distinct gain to individual psychology.

The student and physician must enter more fully into those problems than can the writer in the space allotted. The presence of instinctive reactions to fear of every kind is present in every one. With the child, their unhampered expression is not regarded as unnatural. With the adult, however, shame and pride offer often a sufficient reason for them to seek to repress their presence. Instincts are not unreasoning, even if automatic, but are the accumulated neural experiences inherited from our ancestors. They must be given expression and not repressed. Every trauma which brings out emotional responses would produce no long-continued effect if the individual realizes their real nature and rationalizes correctly concerning them. That is, not only the traumatic situation is thereby correctly appreciated, but also its emotional effects. This is what occurs in sublimation, a term often used in present day literature on the subject.

The symptoms of the traumatic neuroses are largely hysterical in type and, therefore, many of the explanations conform to those of hysteria in general. They deal with various phenomena in a more or less positive way without being searching enough in their basal conceptions to be entirely satisfactory.

Babinski¹ lays down the dictum that hysteria is a psychic disorder whose basal phenomena are due to suggestion (autohypnosis) and to be cured, therefore, by persuasion. He prefers, therefore, to call it pithiatism. He separates from hysteria the effects of emotion upon the endocrine organs and the autonomic nervous system. He has recently reaffirmed his viewpoint based upon his war-experiences. Many observers of the war-neuroses are inclined to accept this hypothesis of Babinski, looking upon the emotional shock as a determining cause, which, followed by an intermediate phase of meditation (Charcot), is succeeded by the symptoms of suggestion. As Dupré put it some years ago, "L'emotion, etant un element de dislocation de la personnalité, favorise la suggestion." Pearce Bailey,⁴ in a recent discussion of the war-neuroses, accepts Babinski's conception of hysteria and notes that the disciplinary control and punishment which were permissible under

army regulations served to prove the efficiency of this method of approach and its consequent cure. "Such a cure," he says, "is quick and pragmatic and adapted to be applied to large numbers, as opposed to being time-consuming and analytic and individualistic."

The effect of ideas upon bodily function called attention to by Möbius, the exaggerated emotivity of Vogt, the narrowing of the field of consciousness of Janet, all served to emphasize various psychological factors present in hysteria. Other attempts at an explanation are pretentiously involved but do not furnish much of value. Such are Hellpach's definition of hysteria as an intensive, extensive and qualitative increase of psychogeniophysical activities and their disproportion to the emotional moments involved; Kohnstamm's conception of a defective idea of health as a basal hysteria phenomena; and Binswanger's opinion that ideas, feeling states and sensations in hysteria have an abnormally increased influence upon cortical and infracortical mechanisms, resulting at times in inhibition, and at other times in over-active responses.

Strümpell and others have emphasized that the desire for compensation is the chief moment in keeping up the emotional results of an accident and thereby "fixing" a neurosis. No difference of opinion can exist that this factor often is the dominant one. Associated with this, is the dread of economic loss by reason of possible disability; a resultant, too-intense self-appraisalment (over-personalization) with the secondary production of a depressed state and irritability (hypochondriasis). Traumatic neuroses, however, occur also in those who do not seek, or have no expectations of receiving, damages. The wish to remain ill must always be considered in litigation-neuroses and the end of litigation often, though not always, brings out a determination to get well. Similarly the signing of the armistice rapidly "cured" many psychoneuroses in our army camps. In the same way, preëmpatory commands after explanations cured many hysteric soldiers in the line hospitals, persuasion in these cases being reënforced by fear of ridicule and of reproachful internment. Psychic contagion as a causal factor and mass-suggestion in the cure were potent psychic factors in war-hysteria. Nevertheless the other war-psychoneuroses did not respond to suggestion and reëducation as did those of a hysteric type.

Diagnosis.—Every patient must be subjected to a complete examination. A traumatic neurosis can only be a diagnosis by exclusion. There should be no occasion for observations like that of Zweig, who reported in 1908 five out of six injuries to the vertebral column falsely regarded as "traumatic neuroses," all because of the neglect of having skiagrams made.

Bárány's tests, in conjunction with other modern diagnostic methods, help to eliminate from the traumatic neuroses many of the hitherto doubtful types. Whenever actual organic symptoms are found, a traumatic neurosis syndrome cannot be the diagnosis.

Inasmuch as diverse social and legal problems center around this diagnosis, the importance of eliminating simulation is obvious. The subjective symptoms, even if they answer to those of the neuroses, should

be corroborated by *objective signs* of disordered function. It is unfortunate that the antecedent medical history of the traumatic neuroses is rarely known or ascertainable. A searching for objective evidence of disordered function must be thorough. Claude⁸ claims to have frequently found in many of the war-neuroses alterations of pressure in the cerebrospinal fluid and adds that "il est possible que les troubles fonctionnels observés soient entretenus par des modifications légères dans la constitution et la circulation du liquide céphalo-rachidien." Gierlich at the 1918 meeting of the German Neurological Association claims that among 230 neurosis cases, 80 per cent. had a lymphocytosis and a neutropenia (Kocher's blood-picture). He concludes that this blood-picture corresponding to that of a ten to twelve-year-old child is the result of an infantilism of the blood-producing organs and is an important objective sign in the diagnosis of all neuroses.

The *objective signs* of a neurasthenic type which should be searched for are: (1) exaggerated tendon reflexes; (2) increased mechanical excitability of muscles and nerves; (3) fibrillary tremor; (4) fatigability; (5) abnormal irritability of the cardiovascular system; (6) insomnia; (7) loss of weight. Insomnia can be confirmed as actually existing by competent observation. Loss of weight generally accompanies loss of sleep. Fatigue offers two methods of objective approach: (a) on the mental side by Kraepelin's method. This consists in having the patient carry-on a series of simple calculations, noting the increased number of errors and the delay in time for their accomplishment as the test proceeds. Accompanying this mental effort, various physical factors of fatigue and anxiety appear, such as excessive perspiration, cerebral congestion, tachycardia and fibrillary tremors. (b) On the physical side, the ergographic method of Mosso is valuable. The curve invariably convex in healthy subjects is straight or concave in hysterical individuals.²²

As regards the *cardiovascular signs* of irritability, tachycardia and arrhythmia without organic evidences of heart-disease, are common symptoms. Erben has called attention to the slowing of the pulse about five seconds after crouching with knees bent and head and trunk flexed forwards. Orthostatic tachycardia—the pulse-rate accelerating markedly upon rising, after having the patient lie down—is often elicited. The "effort syndrome," called D.A.H. (disordered action of the heart), is a picture closely allied to the psychoneuroses in which cardiovascular symptoms predominate. In reality, it is a syndrome of disorder of the autonomic nervous system and is akin, therefore, to the traumatic neuroses. C. Macfie Campbell⁹ has called attention, therefore, to the necessity of a psychopathologic examination as well as one of the heart's mechanism in such syndromes. He presents the problem as follows: "(1) What is the intimate mechanism of the cardiac inferiority? (2) What are the more complex factors, emotional and situational, that precipitate and foster the symptoms? The purpose of the examination is to determine: (a) whether constitutional factors have played a part in the sickness; (b) whether past experiences have conditioned the

present reactions of the patient in a morbid way and (c) whether present instinctive conflicts or a personal situation of much emotional value may not be contributory causal factors."

From the *symptomatic* side, slight cardiac disorders require a close analysis of other etiologic factors not directly due to the accident, especially excessive smoking, chronic alcoholism and syphilis. Traumatic myocarditis is also difficult to judge in reference to the actual strain incurred in an accident. That trauma may produce arteriosclerosis is accepted by competent observers. The cerebral symptom-complex of tachycardia and vertigo, accompanied by a rise of blood-pressure and congestion of the face and conjunctiva, is frequently found after head-injuries without any focal lesions or fracture of the cranial bones. Oppenheim has recorded the presence of an exophthalmos upon stooping accompanying this syndrome. Constant headache bears a different aspect if it is accompanied by flushing of the face and pronounced tachycardia. A vertigo needs to be investigated in the same way; there should be a searching for fear factors (phobias), accompanied by disturbances of the vegetative nervous system; and a careful testing of the vestibular apparatus by caloric and rotary tests should never be neglected.

The complaint of *deafness* is a frequent one in the traumatic neuroses. If it actually exists, it is important to ascertain whether it was of longer standing than the accident. It is not so difficult to decide whether organic lesions are responsible as it is to eliminate hysterical from simulated deafness. Monocular *blindness* requires also a close inquiry into its actuality, organic or hysterical, or the possibility of its being simulated. The tests for special sense examinations are multitudinous. They cannot be entered into here.

The subjective complaints of a *sensory* character—pains, burning and creeping sensations, etc.—cannot be confirmed objectively. Mannkopf's method of a rise in pulse-rate consequent to pressure upon a painful area is not trustworthy. A well-coached subject may claim a hyperesthesia in an area similar to that innervated by a nerve or a cord segment; or he may present the patchy or physiological sensory hyperesthesia of hysteria combined with anesthesia for some stimuli, and changes upon distracting the attention may also be simulated. Analgesia can be easily simulated unless opportunity is found to test for it under positive assurance that the patient is unaware of the examiner's intention.

The electric current is often employed as a diagnosis test of hyperesthesia, utilizing both the lowered threshold for pain stimuli in genuine hyperesthesia as also the opportunity to uncover malingering by pretending in testing to pass a current through a coil. Care in interpreting the answers is necessary as an anxiety-neurosis may be actually present. In the traumatic neuroses, the reaction of the heart-sounds, both qualitatively and quantitatively, to electrical stimulation is advocated by Egger as an objective test in hyperesthesia. In the writer's estimation it is of no value.

Wherever we have hyperesthesia of the skin and pain upon pressure, without any conditional reactions to pain, if we can exclude inflamma-

tory disease, we can be sure we are dealing with malingering. Pressure upon a painful area may, however, produce flushing of the same side of the face (Oppenheim's sign) and may occasionally be accompanied by a dilatation of the pupil with an increased respiratory and cardiac rate (Bechterew's sign), and cannot therefore be regarded as simulated. A dilatation of the pupil upon pinching the skin of the neck or upon pressure over a painful area or upon pricking with a needle (Cramer's test) occasionally helps in establishing a diagnosis, but the writer does not find these tests very reliable.

In mapping out sensory zones, one must bear in mind that it is not a purely objective test. Excluding false answers and interpreting the others in the light of hysteric or organic involvement, we have an objective result, but not till then. Absolute uniformity in response upon different days excludes a neurosis, also a normal innervation, because of the factors of attentiveness, intelligence, warmth and moisture of the skin, fatigue and the varying thickness of the outer skin-layer as well as its blood supply. Müller's method of testing for decreased or lost sensation by varying our examination from above to below the area, and vice versa, often shows the hysterical or simulated nature of the symptom.

Culpin's zigzag test is a better and more reliable method for the limbs. He advocates marking the upper limit and then working from the periphery upwards in a zigzag. The upper limit will shift downwards an inch or so at each test. In both of these tests we are utilizing our knowledge of the difficulty experienced by persons in localizing sensation, or want of sensation, and their necessary errors in interpretation, if it is a functional alteration or a simulated one.

The use of "yes" or "no" in telling the examiner whether he feels or not, often unmasks a simulator who will frequently answer "no" after being touched over a supposed anesthetic area. Stroking two similar areas simultaneously, interspersed with a quick change from the anesthetic or hyperesthetic areas, now on both, or only on one side, is a useful method to employ. The reactions over "ticklish" areas, the reactions of the mucous reflexes, the variations in time of the sensory reactions to stimuli upon the two sides of the body, are important aids in establishing our diagnosis. The influence of psychic factors must always be kept in mind in hysterical analgesia. We must remember its association with hyperesthesia (the production of defense-movements, if we can succeed in suddenly shocking or frightening the patient), and likewise, the associated motor symptoms. Zonal analgesia is not difficult to map out in order to determine its genuineness or not, but a simulated analgesia of the entire body is very difficult to test. [The author is reminded of this because he has at present under observation a man with an analgesia of the entire body.] Heavy electric currents must be employed in such persons, if malingering is suspected. To differentiate an organic involvement from a hysterical one is not difficult. To distinguish a simulator from a hysterical person much ingenuity is necessary, as both may present the same bizarre and unphysiological

reactions. In fact, according to the writer's point of view, *malinger*ing is a protective reaction and hardly possible in the average normal person. The difference lies medically upon the factor of being conscious or aware of the situation and legally upon the credulity and credibility of both patient and physician. The association of exaggeration and hysteria must not be ignored. For a careful analysis of various tests, works like that of Collie,⁹ and of Jones and Llewellyn²² must be consulted.

With the *motor phenomena* of the traumatic neuroses and their diagnosis we have already dealt. We could call attention to the necessity of the general signs of neurosis being present in order to decide absolutely doubtful spasms and contractures of a local nature. Habit spasms or habit palsies are very common after traumatisms. Habit palsies are due to pain, set gaits, postures, or attitudes being indulged in to relieve the person from the using of injured muscles and joints; the position of the body or limbs becomes fixed and habitual, long after the pain has gone.

The differences between true and false paralysis involve a close examination for signs of organic involvement, coupled with a searching for behavioristic reactions which reveal *malinger*ing. The use of drugs to produce changes of the reflexes, of ligation to effect local signs of disease, of the local use of sugar to "doctor" the urine, and the chewing of red dyes to simulate hemorrhage, are occasional methods employed, coupled with various subjective symptoms to simulate a traumatic neurosis. Such combinations rarely, however, give the examiner any trouble in their detection. Nevertheless when such experienced observers as Pierre Marie and J. Babinski speak of their difficulties in separating fraud from hysteria, one does not wonder that errors are made between fraud and the traumatic neuroses in civil life, where the opportunities of the military officer do not exist, and but one examination is often all that is permitted the physician. Rapid recovery is not indicative of fraud, if we were dealing with a hysteric paralysis. Statistics are, therefore, of no value except we analyze not only the symptoms together with the type of accident, but also the after-results of monetary settlements.

We find that a seemingly severe hysterical neurosis soon disappears after compensation is arranged for. On the other hand, the hypochondriacal-querulous types and those with a cerebral or vasomotor symptom-complex are not so easily influenced by a monetary settlement. Exaggeration of actual symptoms exists in 90 per cent. of all psychoneuroses. Simulation or pure *malinger*ing is common enough and always to be looked for. At least 25 per cent. of the traumatic neuroses which get into our courts are pure simulation.

Treatment.—A. GENERAL TREATMENT.—General therapeutic efforts in the traumatic neuroses are directed to: (1) treatment of the surgical results of the injury; (2) treatment of the physiological results of the injury; (3) treatment of the after-effects of the accident involved in the general problems of functional decrease in working ability, financial worries and litigation possibilities, as well as the medical symptoms.

The immediate treatment, after first-aid services, is directed, therefore, to an estimation of how badly the nervous system is injured. Coming by exclusion to a diagnosis of a functional disorder or neurosis, we direct our efforts to combat the effects of fear and fright, and to suggest a reasonable mental attitude in the situation until adjustment is secured and the vegetative nervous system is quieted. **Sedatives and hypnotics, isolation and careful nursing**, are all proper adjuvants at this stage, if care is taken that the reasons for their use are not misunderstood. Often in no other way can the vivid memory-pictures of the horrors of the accident be made to subside with their production of fright-dreams and insomnia resulting from them, together with its train of physical disorders.

When confronted with actual hysterical paralyses or hypochondriac ideas, **psychotherapy** is imperatively necessary. Isolation, forceful suggestion and reëducation are the measures needful. These therapeutic aids must be used without equivocation or compromise and must be intensively applied. Their success often depends upon a quick response on the part of the patient because in no other way can doubt be dissipated. An early return to work often helps to "fix the cure," although the physician must be careful in estimating the working ability of his patient before returning him to work.

An instructive lesson in the treatment of traumatic hysteria was offered by the recent war. In the first place, the French secured brilliant results, as far as removing symptoms and sending soldiers back to the front were concerned, by an energetic use of electricity supplied through long connecting wires, carried on an overhead trolley the entire length of a room. They call this *torpillage*.⁴ The war neuroses are not pensionable in France. This has been a great aid to their suggestive discipline. The author has repeatedly found the isolation room as introduced by Déjerine of great help in breaking down many of the bad mental traits of hysteric patients. Persuasion in civil practice can only reach the plane of discipline by its use; and it must be remembered that the absolute control of a patient, whether a war hysteria or a traumatic hysteria of civil life, is necessary. This is discussed more fully later.

The readjustment of the thoughts and feelings of cases of chronic traumatic neuroses of long standing is not as uniformly successful as with recent types. An analysis of their mental life is generally necessary to effect a permanent cure. This is even more vital when the neurosis is an obsessive or compulsion type, partaking of the nature of what is understood by psychasthenia. **Physiotherapy or ergotherapy** is here a necessary adjuvant to psychotherapy. Pearce Bailey expresses a very important point in emphasizing that the resumption of duties tend to build up character. Therapeutic workshops should be established in every industrial center.

B. TREATMENT OF THE HYSTERIC MANIFESTATIONS.—The large experience offered by the war has shown anew that traumatic hysteria is an eminently curable condition. It has likewise proven that results are better obtainable through quick intensive methods than by temporizing,

once a diagnosis has been established. One sitting has usually sufficed for a removal of the symptoms. Military discipline stepped in to utilize the suggestibility of those soldiers having a traumatic neurosis, the physician explaining the reason for the disability, suggesting motion in the non-used limb, utilizing electricity to reënforce his appeal, threatening disclosure and punishment when necessary in recalcitrant subjects, and dominating by force of personality the mind of the subject. The first evidence of a return of normal function was seized upon to reënforce previous persuasion, and the patient was not left until he was entirely relieved. The three factors of success were: (1) the detailed examination completely establishing the presence of hysteria; (2) a physician of understanding, tact and determination, without bluster or bullying; and (3) the utilization of intensive psychotherapy at the opportune time. The latter is the new point of departure in treatment. Patients were rested who needed it; others were isolated, if necessary; still others were placed where they could behold the betterment of other similarly affected. But when the time for treatment came, this was pushed to an issue, no matter how many hours it required. Cases of mutism or aphonia, monoplegia or diplegia, deafness or blindness, in countless numbers, were taken in hand in this way, and relieved of their symptoms.

Yealland,³² Purves Stewart,²⁰ Colin Russell,²⁷ Wolfsohn and Farrar¹⁵ are some of the English and American officers who have followed Babin-ski and Roussy and others of France in this method. The hysteria of the war offered a more hopeful expectation of betterment than the traumatic hysteria of civil life, even admitting in the cases of the fighters some predisposition to a neurosis as evidenced by the frequency of nervous disorders in themselves or relatives in the previous history. The suggestive factors were more obvious. The strivings and fears of the soldier were easily ascertainable. The environment for treatment could be more carefully selected. No adverse influences on the part of friends and relatives existed. Not many were "hospital-birds," which name was given to those who had wandered from one institution to another. Few had the impression of serious disability confirmed as in civil life by suggestions of spinal disease or by splints put on for the correction of the contractures. Purves Stewart²⁰ warns with regard to this that "splints and other apparatus, originally applied for the maintenance of correct posture in a limb which has some *bona fide* organic lesion of nerves or other structures may subsequently come to exercise a baneful suggestive effect, so that by the time the surgical injury is healed, the patient has lost the habit of using his muscles. In other words, a hysterical paralysis has become superimposed on an organic injury." [Recently a railroad fireman was sent to the author for diagnosis and treatment because of the loss of the use of his right arm as the result of an accident. Inasmuch as the examination revealed the non-organic nature of the injury, the writer utilized faradism and explained away the paralysis. A week later, the man again came to the consulting room with the same disability. In the interval he had gone to his family physician who had applied an arm-splint. By reënforcing the

former suggestions and showing why no splint was necessary, the patient went away "recovered," and stayed well.]

The treatment of hysteria among the soldiers was an early and a brilliant success in the French Army, then, because they not only excelled in psychotherapy, but they early instituted measures to counteract adverse suggestion. In 1916, the French neurologists established the following rules:

1. For hysteric symptoms, neither discharge from service, pension nor gratuity will be offered.

2. Whenever hysteric symptoms are associated with organic affections, no account of the hysteric symptoms is to be taken in estimating the degree of incapacity and, therefore, the patient's pensionable disability.

Later the English and Canadian forces adopted similar measures, with corresponding beneficial results. The counter-idea-to-betterment from compensation or pension was thus removed.

The treatment is not always as simple as outlined above. A general **anesthetic** occasionally is advisable to overcome contractures or to give opportunity for suggestions to stubborn patients while coming out from its influence. Like the use of hypnosis, such measures are, however, rarely necessary. Eder, Smith, Pears, and Kennedy are some of the English and American war reporters whose treatment, equally successful to those who were cured by a rapid-intensive method, is regarded by some as having insured greater durability of the cure by having been effected through psychic analysis.

The realization of various trends mutually antagonistic influencing our emotional life is generally accepted to-day. We recognize the fallacy of the James-Lange theory concerning our emotions, in that they failed to recognize that emotions are forces which are at hand to be used unconsciously and consciously, and that they, therefore, primarily influence behavior, as well as cause physical reactions in the body (Shand²⁸). Why the angry man becomes more erect or the frightened man pales and trembles cannot be explained without reference to our primitive instincts. Upon their integrity is emotional adjustment dependent. And so, that treatment is often best which can reason out the fundamental sources of the hysteric disability and utilize them in explaining away the symptoms, rather than the one which gives only a physiological explanation. It may be remarked *pari passu* that neither may always be correct; and the explanation given may be modifiable by a future increment to the physician's knowledge. Mutism, for instance, is explained by some authors as a compromise between repression and inhibition. Russell²⁷ analyzes mutism, and reveals another method of approach by calling attention to: (1) the close association between fear and the voice centers; (2) the failure of the voice by reason of spasm (over-stimulation) during intense fear; (3) the fear having passed, the memory of it remains; (4) the suppression of this memory entails the non-use of the voice-mechanism, because of the association set up between the voice center and the center of fear. [A loud

cry of alarm was heard by a traumatic neurosis patient¹ of the author's at the same instant that a piece of metal fell from the roof of a steel mill he was inspecting in the course of erection; it knocked his hat off, scuffed the back of his overcoat, and buried itself a foot in the ground. From that day he has never been able to walk under any constructing work and when necessary to do so, orders all work to cease for the time being. Yet this situational fear and its emotional reaction were apparently forgotten for five years, the time which elapsed between this accident and his coming to the author on account of an anxiety-neurosis.] The elucidation of emotional upheavals in the lives of our patients forms the subject-matter of a special method of treatment, called psychic analysis, and which, under the name of psycho-analysis, has a literature of its own so large that we cannot enter into it here. Many regard involved psycho-analysis in the Freudian sense as a waste of time. Our analysis must be just as involved as is necessary in order to obtain a psychic cause. [In the case of a patient whom the writer recently cured of an hysterical tic of the neck-muscles, had he not obtained by these methods her confession of having been raped by a boy friend when she was a child of twelve and her mental conflict from that time on, she would not have been cured.] Often short-cuts serve our purpose; at times, weeks must be spent in these preliminary findings in order to uncover the "memory-seal"¹³ (Deckerinnerung of Freud).

No matter how recovery is secured, it should never be exploited. Nor should the physician lessen his dignity and accept exaggerated congratulations from the patient or family. The threshold of elation is bound to change and close upon the physician adversely should he be such a poor psychotherapist as to allow himself to be classed with miracle-workers. Successful psychotherapy implies, on the part of the physician, confidence in himself based upon knowledge and skill. To get at all possible sources of emotional upsets requires a close insight into psycho-analysis, secured only by its practice. And so dream problems and other symbolic representations must occasionally be studied even though we do not accept any fixed interpretation. The traumatic neuroses of the war were considered in this way by Eder. He carried out with success strictly Freudian methods in his treatment. Similar results by different methods forces recognition of environment and of the physician's personality as factors quite as important as is the plan of treatment. Even the rough procedure of Kaufmann, the *Ueberrumpelungsmethod*, often succeeded according to German army reports, although occasionally, as Oppenheim reported of one patient in condemning this method, the patient's wrist was dislocated while being held down in order that intense faradic electric currents might be administered. It is interesting to note that Russell used military sternness with some, and "reason, persuasion and encouragement" with others. We agree with him that the blustering, truculent hysteric is generally a malingerer.

C. TREATMENT OF EXHAUSTION AND EFFORT SYNDROMES.—Since we are dealing here with patients in whom the physical factors of fatigue

and irritability dominate the treatment we must adequately combine rest, exercise and hydrotherapy with the psychotherapy needed. The physician must endeavor also to terminate all legal worries as soon as possible. He must try to nullify the ill-effects of economic difficulties before he attempts to further drain the resources of his patient. So many patients of this type are of low intelligence. If so, do not expect to remake them and do not try, as many physicians often do, to put in their hands books upon psychotherapy which they cannot understand. Books are at the best poor makeshifts for what the physician should be able to impart verbally.

The psychasthenic states are never the result of accidents. These patients were sufferers from a neurosis before the trauma. If attempts to obtain compensation for such symptoms are made, it could only be from claims of an exacerbation of symptoms. Treatment of the psychasthenic states are tedious and not as brilliant as in hysteria. But psychotherapy will, if consistently carried out, produce at least a pronounced betterment. With the routine advice so often given of climatic cures, the writer has no patience.

D. TREATMENT OF OTHER SYNDROMES.—The treatment of the other syndromes differs according to the type of symptoms. Local irritation from scar tissue requires surgical intervention. Evidence of thyroid intoxication requires rest and perhaps eventually ligation of the thyroid vessels, independently of the question of a primary or secondary Graves' disease. Epilepsy after trauma generally demands surgical attention. Concussional types of the character of Friedman's symptom-complex need rest and avoidance of work and worry for a prolonged period. The amnesia of trauma, if of a functional type, is amenable to the same plan of treatment outlined under traumatic hysteria. A state of negativism with Ganser's symptom of irrelevant answers is frequently hysterical; curable, therefore, if the physician does not regard it as indicative of dementia præcox and avoids appropriate psychotherapy.

More than anything else in treatment a frank and honest stand by physicians is required to counteract the suggestive influences clustering around injuries and the compensating features of our laws. The suggestion to the physician of fees secured by helping in establishing an adequate claim for damages should be fought against. The physician's aim should be to cure uninfluenced by any medico-legal considerations.

E. PREVENTION OF TRAUMATIC NEUROSES.—This requires a consideration of means to raise the social position of workmen, to relieve their economic stress, to better our methods of compensation and to educate children in respect to self-control and the influence of the emotions upon bodily functioning. The physical training of workmen, avoidance of alcohol and efficient medical handling of those who have been in accidents, are measures of prevention which soon will be generally adopted. A survey of those engaged in hazardous work in order to eliminate the unstable or the predisposed is an ideal which is probably unrealizable.

Prognosis.—The traumatic neuroses of the war have had an unusually high recoverability rate: (1) In the French Army from the beginning;

(2) in all Allied Armies in the latter part of the war. This is accounted for by: (a) the average youthfulness and general good health of the soldier; (b) the preponderance of traumatic hysteria among the neuroses of the war; (c) the influence of the military command in suggesting recovery to the sick soldier, coupled with their treatment by neurological divisions among conditions conducive to betterment. The French Army had a better recovery rate from the beginning, because with them a traumatic hysteria is not a pensionable disability, as it was at first among the other armies. We shall again speak of this later. Even in Germany, where under their compensation-system of civil life hopeless prognoses were formerly more the rule than elsewhere, the war has made them take a more hopeful view. The incidence of recoverability depends in civil life upon many diverse influences:

1. A competent opinion and the actual existence of a traumatic neurosis uncomplicated by the presence of an organic disease.

2. The comparative age of the traumatized person together with fairly good habits as regards the use of alcohol, tobacco, sleep, etc.

3. The environmental factors of social standing, economic position and the presence or absence of litigation in connection with the patient.

4. The conception of recoverability in connection with: (a) removal of all symptoms, (b) a return to work or comparative usefulness.

Traumatic hysteria is obviously of a better prognosis than is a traumatic neurosis with pronounced hypochondriasis and phobias. The intensity of reaction on the part of the vegetative nervous system must be interpreted from the standpoint of the constitutional makeup of the individual, the resultant disturbance in function of the endocrine glands, and the intensity of the emotional or psychic upheaval. And prolonged litigation, desire to avoid war-risks, chances for life pensions, and, in civil life, the attitude of friends and family are all factors of importance in estimating recoverability. The habit of avoiding all work to secure a better adjustment of claims is in itself a bad prognostic feature if the individual is neuropathic by inheritance, or elderly.

Naegeli's statistics showed that 93 per cent. of the traumatic neuroses returned to work after an adjustment of their claims. Brand claimed that in Austria the traumatic neuroses increased greatly after their pension law went into effect. The non-recording of such a syndrome before the inauguration of laws for compensation does not, however, prove that it did not exist. Stierlin,⁴¹ three months after the Messina earthquake, examined 226 victims and found that 20 per cent. of them had objective signs of the traumatic neuroses; yet, immediately after this catastrophe, Bianchi reported that from among 600 survivors not one had signs of a neurosis. A period of latency (contemplation, expectation) between an accident and the production of psychogenetic symptoms is often noted. The immediate appearance of the traumatic neuroses may be regarded as being due to their presence before the accident, unrecognized or suppressed. The anticipatory fear accompanying the trials of trench life have been described many times. The necessities of earning a living likewise in civil life often makes an individual suppress his neurosis until

an accident removes all barriers to its expression and detection. Dercum, some years ago, followed Babinski in including only hysteria under the traumatic neuroses; he claimed that when litigation was not a factor, the symptoms disappeared readily under the influence of suggestion, unless the attitude of the patient's physician confirmed the invalid in his belief of injury. The rarity of treatment of the traumatic neuroses after litigation is over is a common enough observation. The results of the treatment of traumatic hysteria in our armies in France were favorable in about 90 per cent. of the cases. There is a unanimity of opinion that the pension or annuity systems of compensation are not conducive to recoverability. Likewise, delay in litigation serve to "fix" the neurosis and hinders betterment. A too early return to work before the fear-reactions are over often serves also to influence the idea of non-recoverability, and thereby to increase the depression and to influence the emotions of the victim of an accident.

We must always be guarded in our prognosis of the cerebral or vasomotor symptom-complexes, especially concussion types with a lacunar amnesia and a persistent vertigo. Electrical accidents have generally a poor prognosis, inasmuch as generally some organic involvement is present whose presence must be carefully searched for. They may, however, be entirely psychogenetic in type. The possibility of a psychosis developing from an accident-neurosis is always to be kept in mind, even though it is not of frequent occurrence.

The prognosis of the traumatic neuroses depends, therefore, upon various causative factors: the nature of the syndrome present, the social conditions of the patient, the legal factors involved in the settlement of claims for damages and the constitutional (psychological) makeup of the individual.

Medicolegal Considerations.—The right to recover damages for injury due to the negligence of others is incontestable. The increase of industrial machinery and the growth of rapid transportation have filled our courts with suits to recover enormous damages for minor injuries. Street railway corporations yearly pay out from 5 to 10 per cent. of their gross receipts in settlement of claims growing out of accidents. The traumatic neuroses naturally, therefore, are more frequently defended in the courts than other syndromes. Many attorneys and a few physicians devote practically all their time to this kind of work. Unconscious malingering, manufactured symptoms, exaggeration of those present, or false interpretations of them, are daily encountered in our courts, and unjust awards are often given by a bewildered jury. Angell has said of "railway spine": "It is a convenient and picturesque term which has hypnotized juries even as the shock has hypnotized the plaintiff." The following points stand out:

1. Our method of jury trials in which biased opinions carry as much weight as do more careful, if not more competent ones, are unfair. How to remedy it is not, however, clear. A jury of physicians is unobtainable; and even if it were, such a jury would simply multiply the number of physicians in the employ of legal firms and corporations, and the better

element of the profession could not be induced to serve. A commission of physicians to report to the judge, whose charge to the jury would be based on their findings, would not secure any betterment. The author is convinced that a commission on which each side has appointed a representative, and the court a third member, also fails for obvious reasons to give good service. As Sir James Stephen, Y. C., says in his "History of Criminal Law," "The judge and the jury alike are and ought to be instructed only by witnesses publicly testifying in open court on oath."

2. Since our common law permits each side in litigation to secure as many experts as they desire, the legal firms which specialize in accident cases and others, also, by employing physicians on contingent fees, sometimes unduly large, have always at their call physicians who, sharpened by experience, give their testimony the proper bias to influence the jury. This conspiracy of interests is reprehensible.

A change of law, which would give only the court the right to call experts is impossible to secure, and even if brought about, would not prevent the use of politics by those interested in securing the appointment of the "right kind" of physician. If it were possible to create a law which would have the compensation of all experts a matter of court record, by making it a part of the expenses of the trial, some improvement over present methods would, however, be attained.

3. Jury awards with us seem to place a premium on exaggeration, and a proper staging of the dramatic possibilities of the client's case secures an increased verdict. Awaiting for years a settlement often nullifies all efforts to bring a claimant for damages back to usefulness, by reason of the psychic effects of not desiring health, together with the bringing on of postural spasms and contractures through fixed positions. The German system of an indemnity of two-thirds of a person's wages after the fourteenth week does not remove these factors. The Danish system of a cash settlement is different from ours in that physicians, not juries, judge the matter; they accord the claimant a small daily amount until nine months have elapsed, when they decide on his compensation, and one and a half years later give a further and final settlement, if he has not recovered.

4. One great suggestive influence in litigation is the blanket indictment of complaints introduced by the attorney. The averment in it that John Doe is a hopeless nervous wreck is read by the plaintiff before he signs it, and the coöperation of a physician who agrees to the truth of such a statement helps to fix this idea in the plaintiff's mind.

5. A physician finds great difficulty in securing an opportunity to examine a claimant for damages in a satisfactory manner, if he is not retained by the claimant's side. The examination may be requested at the attorney's office in the presence of a "runner" who coöperates in the examination. The plaintiff's physician is utilized to suggest both to the plaintiff and to his colleague; and a second examination is often refused. Sir John Collie did a great service in England when he insisted that he be given the opportunity for examination without the

presence of "runners" and friends, and, whenever possible, at his office, where he had at hand his instruments of precision. He secured a legal decision, which was affirmed by the higher courts, that a "solicitor's office is not in ordinary circumstances a proper place at which to hold a medical examination of a workman."

6. Medicolegal opinions should never be based on subjective symptoms only, and objective symptoms must be present also in order to establish a diagnosis in court. The subjective symptoms are not based on actual facts. They are not the findings of the physician, and they must not exercise an undue influence upon the physician. The author has known physicians to base a diagnosis of epilepsy on the spasms and spells of somnambulism that were manufactured for him; or to base a diagnosis on unconsciousness supposed to have followed an accident which later was shown never to have taken place, or to diagnose "brain-abscess from trauma" from a subjective vertigo, head pain, and a bradycardia which was familial. Such mistakes in judgment would be avoidable if the physician dealt more with facts.

7. Hypothetical questions will continue to be given. Distorted medical facts, exclusion of important symptoms, and over-emphasis of minor ones make them often of no value. Because the basis for legal redress is a physical injury, slight injuries and their effect are always given undue importance in court.

If the physician has examined the injured person, he should refuse to answer any hypothetical questions, unless they included his objective findings, upon which his answer should be predicated. For him, in court, subjective symptoms should have the weight of hearsay evidence. If the injured person could be examined, and the physician has not examined him, the court would probably refuse to permit him to render an opinion. The physician should also insist that the symptoms which are the direct result of the accident should be separated from those which are not, or probably are not, due to the trauma. Probabilities and possibilities in prognoses should also be definitely separated in any opinion rendered. The law does not require physicians to assume that medicine is an exact science, and honest physicians often do themselves harm in court by being too positive. Often, however, positive statements dishonestly offered secure a bigger verdict for the claimant. Positive prognosis in traumatic neuroses is generally impossible unless the physician has personal knowledge of all medical, familial and social factors. He should, therefore, never give opinions based on hurried examinations, or with an imperfect knowledge of environmental factors.

The author is aware that the expert is required to accept as true the evidence of the witnesses incorporated in the hypothetical question. But he is permitted to qualify his answer in order to make it plain that he can have no expert opinion on evidence which does not agree with his objective findings, and he should as a rule not be satisfied with categorical answers.

8. No statistics concerning prognosis carry, or should carry, much

weight in court because of the personal equation of the nature of the accident and the social status of the patient, and the different groupings found in statistics and variations in the laws of different countries. The future of each psychoneurotic patient must be determined separately by studying his social habits, age, nature of work, intelligence, emotional complexes, etc., as well as the kind of accident and diagnosis offered. In court, since much of this evidence is not in the hands of the expert, his opinion is to that extent defective. But more defective still is the opinion rendered on the appearance of the patient, subjective symptoms, and the sympathies aroused by the litigation. In general, however, the physician is not truthful who renders an absolute opinion that a traumatic neurosis is not a recoverable disease.

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CHAPTER XXIII

VASOMOTOR AND TROPHIC NEUROSES

BY S. PHILIP GOODHART, PH.B., M.D.

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ANATOMY AND PHYSIOLOGY OF THE SYMPATHETIC AND THE EXTENDED VAGUS SYSTEMS

The influence of the sympathetic and extended vagus nervous systems upon vasomotor and trophic neuroses is now generally conceded. It is, therefore, necessary that we consider the anatomy, physiology, and clinical manifestations in their relation to these two important systems of nerve control.

Sympathetic Nerve.—According to Higier, whose clear conception of this complicated subject is well worthy of study, the *sympathetic nerve* constitutes the vegetative nervous system and essentially controls the mechanical functions of the viscera.

A distinction is made between its trunk and its branches. The trunk, or gangliated cord, is divided into three segments: the cervical, thoracic, and abdominal segments. The branches are likewise divided into three portions: the arterial, peripheral and communicating branches.

SYMPATHETIC NERVE TRUNK.—The trunk or gangliated cord represents a symmetrical organ which passes directly in front of, and parallel with, the vertebral column, taking an extrapleural and an extraperitoneal course from the base of the skull down to the coccyx, usually converging at the lower end in a loop, a filament or a single ganglion.

In certain of the lower animals (fishes) which have a strictly segmentary structure, the gangliated cord possesses a ganglion corresponding to each vertebra or spinal segment, so that it conveys the impression of a chain interspersed with ganglia, resembling a string of beads.

Each *sympathetic ganglion* usually lies on its vertebra, and is therefore known as the sympathetic or *vertebral ganglion*, as distinguished from the *spinal ganglion* which belongs to the cerebrospinal system, clings to the posterior sensory root and, because of its anatomical position at the intervertebral foramen, is named the spinal or *intervertebral ganglion*.

The sympathetic ganglionic system is best marked in man at the thoracic and upper lumbar segment, where the segmental-metameric type is relatively well preserved, so that twelve sympathetic ganglia correspond bilaterally to the twelve thoracic vertebræ and ribs.

In the cervical and sacral segment, where the embryonic type has disappeared, the ganglia sometimes become confluent, as may be recognized from their mulberry-shaped surface. At the neck we accordingly find only three fused ganglionic conglomerates—the upper, middle and lower cervical ganglia. At the lumbar segment, several weak, incomplete ganglia are found.

This practically represents our knowledge of the sympathetic trunk, the vertebral ganglia of the sympathetic gangliated cord, and their relation to the spinal ganglia of the cord.

SYMPATHETIC NERVE BRANCHES.—In describing the sympathetic branches, a distinction is usually made between the following:

1. *Arterial Branches, or Vascular Plexuses.*—These are:

(a) *Head plexus* or *carotid plexus*, which begins at the upper cervical ganglion, passes toward the skull, surrounds the carotids, and supplies the cranial cavity with sympathetic fibers.

(b) *Chest plexus* or *aortic thoracic plexus*, for the heart, aorta, lungs, and esophagus.

(c) *Ventral plexus* or *aortic abdominal plexus*, which surrounds the three large branches of the abdominal aorta and supplies the abdominal organs and the mesentery with sympathetic fibers.

(d) *Other smaller plexuses* are: the laryngeal, thyroid, cardiac, pulmonary, esophageal, celiac, mesenteric, renal, spermatic, hypogastric, uterine, vesical, and cavernous plexus.

2. *Peripheral Branches.*—The peripheral branches are:

(a) The *cardiac rami* of the thoracic cavity, which are especially important, are given off from the three cervical ganglia and form the *cardiac plexus*.

(b) The *splanchnic rami* are formed by the six lower thoracic ganglia and pass from the thoracic cavity into the abdominal cavity, where they innervate the gastro-intestinal tract with its appendages.

3. *Communicating Branches.*—These, in their capacity as connecting paths from the sympathetic ganglia to the anterior spinal roots, represent an important avenue of communication between the cerebrospinal axis and the sympathetic system.

Additional Structure of Sympathetic Nervous System.—The sympathetic nervous system comprises, in addition to the sympathetic nerve, a number of large structures, the physiology of which still remains more or less obscure, such as the paraganglia, the chromaffin glandular bodies, the prevertebral celiac, cardiac, and stellate ganglia.

Anatomical and Functional Relation of the Most Important Body Ganglia.—The uppermost ganglion of the gangliated cord, i.e., the *cervical ganglion, superior*, or *sympathicum primum*, receives its pre-cellular fibers from the last cervical segment and from the uppermost dorsal segments. It innervates, besides the blood-vessels, the pilary muscles and skin-glands of the head, the dilator muscle of the pupil, the smooth involuntary orbital muscle of Müller and the post-orbital muscle (of Müller).

The *cervical ganglion, inferior*, gives off—together with the neighboring *first thoracic ganglion*, or *stellate ganglion*—accelerator nerves to the heart (*nervi accelerantes*) and probably vasoconstrictor fibers to the pulmonary vessels.

The largest ganglion of the abdominal cavity, the *celiac ganglion*, has its most important roots at the celiac plexus in the *major* and *minor splanchnic* nerves, derived from branches of the thoracic ganglia. Both leave the chest cavity through an intramuscular gap in the diaphragm and enter the abdominal cavity, where they reach the celiac ganglion as pre-cellular fibers, and then, as mesenteric nerves, supply with sympathetic fibers the gastric glands, the liver, pancreas, spleen, kidneys, suprarenals and intestines down to the descending colon.

The *mesenteric ganglion inferior*, contains pre-cellular fibers from the upper lumbar portion of the spinal cord and sends its non-medullated postganglionic fibers to the colon, and, in the form of the *hypogastric nerves*, to the anus, the bladder, the vesical sphincter, and the genitals.

The *middle dorsolumbar sympathetic segment*, moreover, supplies the end-organs in the skin, the blood-vessels of the skeletal muscles, and the blood-vessels of the viscera between the mouth and rectum, with sympathetic nerve-fibers.

The vegetative nervous system, according to Eppinger and Hess, can be divided anatomically as well as functionally. The first anatomical unit is represented by a system of fibers derived from the middle portion of the thoracic cord and the upper lumbar segment, forming in their further course the gangliated cord of the sympathetic system. After these fibers have passed through the gangliated cord, their anatomical separation becomes very difficult, because the true sympathetic nerves become mixed with others and seldom pass separately to the end-organs.

The second anatomical unit is characterized by the derivation of its fibers in part from the brain and the medulla oblongata, and in part from the sacral cord, without its entering into a relation with the gangliated cord. This system is roughly divided, according to its origin, into a cranial, bulbar and sacral segment. The *cranial* nerve plexus passes mostly through the tracts of the oculomotor nerve, is interrupted in the ciliary ganglion, and supplies certain areas of the eye. The

bulbar segment follows the course of the facial nerve and the glossopharyngeal nerve, and transmits fibers to the glands and vasodilators of the head. Its most important and largest branch is the vagus nerve, the principal nerve of the viscera; it supplies the heart, the bronchial tree, the esophagus, stomach, intestine, and pancreas. The *sacral* branch, known anatomically as the pelvic nerve, innervates the descending colon, the sigmoid flexure, the anus, the bladder, and the genital apparatus.

For the purpose then of brevity, all those nerves which pass through the gangliated cord of the sympathetic nerve are described as *sympathetic autonomic*, whereas all the remaining vegetative fibers are designated as *vagal autonomic* (system of the expanded vagus).

Confusion has arisen because of the lack of uniformity in designation of the two systems—vagal and sympathetic. With Langley we define as the autonomic that part of the nervous system which may be described as independent of the cerebrospinal system in that its nerve-fibers do not go at once to their destination but pass to a ganglion, in which, by a kind of “synapse,” they are broken up and then proceed to the organ which they control. The autonomic system includes the entire sympathetic nervous system—the sympathetic autonomic and certain fibers of the vagus nerve—the vagal autonomic. The two systems are mutually antagonistic.

The anatomical separation of the nerves of both systems is relatively easy in the center and in the vicinity of the cerebrospinal axis, but extremely difficult, or practically impossible, in the intricate network at the periphery.

In the sympathetic as well as in the vagal autonomic system, two kinds of nerve fibers can be distinguished: (a) positive, vaso-viscero-glandulo-motor fibers, and (b) negative, vaso-viscero-glandulo-inhibitory fibers.

The normal state of irritability of the ganglionic cells is safeguarded by the delicate adjustment of stimulation and inhibition, so that the apparently superfluous inhibitory processes must be considered as an indispensable protective mechanism of the central nervous system.

It is another noteworthy peculiarity of the vegetative organs that they are all reached by the sympathetic nerve-plexus which passes through the gangliated cord, and also by the vagal autonomic system, so that there is practically no organ independent of the will which does not possess a double innervation. The only apparent exceptions to this rule seem to be the sweat-glands, the hair-muscles, and the vascular muscles of the viscera, which receive fibers exclusively from the gangliated cord, according to anatomical teachings. But pharmacological tests, which by some observers are regarded as more reliable, indicate double autonomic innervation of these organs, more particularly of the sweat-glands.

This double antagonistic innervation is a factor of enormous importance. Just as the cervicothoracic sympathetic fibers are functionally and pharmacologically antagonistic to the vegetative fibers originating in the skull, i.e., the cranial autonomic system vagal there likewise exists

an antagonism between the thoracolumbar sympathetic nerves and the pelvic nerve, derived from the sacral-autonomic territory (vagal).

In the *midbrain*, the segment of the *oculomotor nerve* which innervates the sphincter of the pupil (miosis), the ciliary body (spasm of accommodation) and, in part, the levator palpebræ (widening of the palpebral fissure) belongs to the vagal autonomic system. The involuntary fibers or Müller's external muscle in the upper lid, are, however, controlled by the sympathetic nerve.

In the *medulla oblongata*, importance is attached to the tract of the *chorda tympani* and the *lacrimal nerve*, which pass to the salivary glands of the mouth and to the lacrimal gland; importance is also attached to the domain of the *vagus nerve*, which passes toward the lung, heart and intestinal canal. It acts on the respiratory tract by contracting the smooth bronchial muscle. The vagus is the inhibitory nerve for the heart, and exerts an inhibitory influence upon the cardiac system in every way, in contradistinction to the sympathetic accelerating nerves. In the upper digestive tract, the vagus contracts the musculature of the esophagus, and of the cardiac and pyloric sphincters; it increases the gastric peristalsis and secretory function. In the small intestine, stimulation of the vagus causes evacuating, rolling movements, more rarely spasm of the bowel-muscle; in the smooth muscle of the gall-bladder and the excretory duct of the pancreas, it gives rise to intermittent spasms. In the pancreas, stimulation of the afferent vagus branches causes a momentary increase of the pancreatic secretion.

Over the entire extent of the *spinal cord*, *autonomous centers* seem to be scattered in addition to the sympathetic centers. These are only demonstrable by pharmacological means. They are connected with the vasomotors of the skin and mucous membranes, the sweat-glands, and the muscles of the hair follicles.

In the *lower segment of the spinal cord* is situated the center of the *autonomic pelvic nerve*, which practically corresponds to a lumbosacral vagus and supplies the descending colon, the sigmoid flexure, the bladder and the genitals.

It is probable that the autonomic nervous system, in view of its relation to the internally secreting glands (pancreas, thyroid), very actively participates in the mechanism of the body metabolism.

With special reference to their *influence on metabolism*, the glandular organs are divided into: (1) accelerative organs (suprarenals, thyroid, hypophysis cerebri), and (2) retarding organs (pancreas, parathyroids, thymus). The first group stimulate the sympathetic nerve, the second inhibit it, states of hyperfunction and hypofunction occurring respectively, in each case. Albumin, carbohydrates, fats, and certain mineral substances, notably calcium, enter into the consideration of this hyper- and hypofunction.

The influence of the endocrine organs upon metabolism has been shown by many observers, including the writer. It is probable that the autonomic nervous system plays the intervening rôle. For example, in cretinism Janney has observed hypoglycemia accompanied by creatin in

the urine. A delay in carbohydrate utilization in a cretin and several exophthalmic goiter patients studied by Janney, was demonstrated by blood-sugar curves. Cushing called attention to hypoglycemia in dyspituitarism and it has also been associated with the pathology of Addison's disease. The relationship between calcium and magnesium retention and output to certain diseases of endocrine origin, has been definitely determined (Janney McCrudden and others). In a clinical and metabolic study of a series of nine cases of muscular dystrophy under



FIG. 1.—PROGRESSIVE MUSCULAR DYSTROPHY OF THE BONES OF THE ARM, SHOWING OSSEOUS TROPHIC CHANGES, RAREFACTION AND UNDERDEVELOPMENT.

observation at Montefiore Hospital, Janney, Isaacson and Goodhart demonstrated the relationship between endocrine dysfunction and the muscular dystrophies. In addition to certain disturbances in the creatinin-creatin metabolism, there was found a constant metabolic hypoglycemia and impaired utilization of carbohydrate. The metabolic findings were practically the same as those recorded in diseases of unquestionable endocrine origin, namely, hypopituitarism, Addison's disease and myxedema. In nearly all of the dystrophy cases of our series, marked vasomotor and trophic changes were demonstrable; trophic changes in hair, nails, skin and bones were observed. Especially interesting and suggestive were the trophic changes in the long bones—rarefaction and underdevelopment. (See Figs. 1 and 2.)

The *trophic action* of the nervous system upon the body metabolism was investigated by Stefani on the basis of animal experiments (on dogs and frogs). He arrived at the conclusion that the nervous system exerts a direct regulatory influence upon the general metabolism, by means of a special kind of nerves which are to be designated as *regulatory nerves* (*vagus*). The influence of the nervous system on the metabolism of the

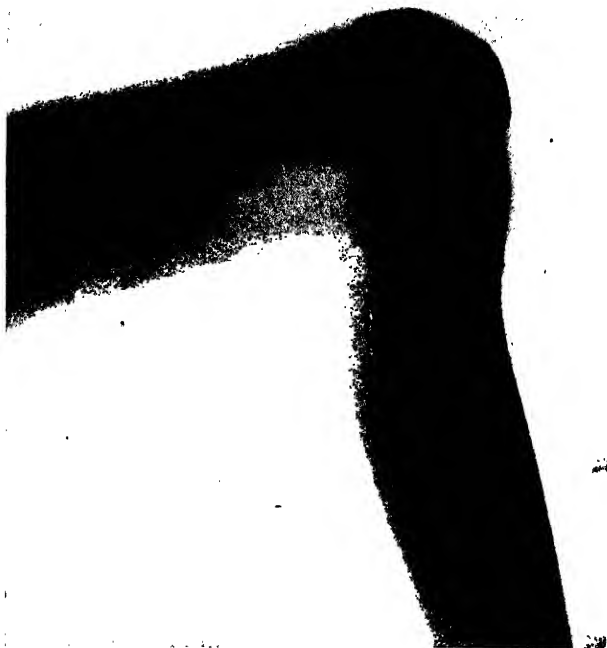


FIG. 2.—PROGRESSIVE MUSCULAR DYSTROPHY OF THE BONES OF THE LEG. (Same Case as Fig. 1.)

individual organs is indirect, in so far as it transmits the functional stimuli to these organs and regulates their blood supply.

The innervation of the large organs of the abdominal cavity is analogous to that of the gastro-intestinal tract. On the liver and pancreas, the vagus exerts a stimulating, and the splanchnic nerve an inhibitory effect. The kidney receives sympathetic vasodilator and secretory stimulating fibers from the lower dorsal roots, but secretory-inhibitory nerves from the vagus.

The functions of the internal organs are controlled by the sympathetic

and vagal autonomic nervous systems. It is by the delicate adjustment of the antagonistic action of these nerves that the visceral tonus is maintained. When the balance is disturbed, various conditions result, depending upon which set of nerves is in the ascendancy. Eppinger and Hess have given the name *vagotonic* to the state in which the vagal autonomic nerves are the stronger, that is, to the state of sympathetic insufficiency, while the reverse condition is called *sympatheticotonic*.

The *sympathetic autonomic system* dilates the pupil, causes the eye to protrude, accelerates the heart action, inhibits intestinal peristalsis, relaxes the anal sphincter, and produces glycosuria and polyuria.

The *vagal autonomic system* contracts the pupil, relaxes the zone of Zinn, depresses the heart action, increases peristalsis, and stimulates the secretion of the gastric and pancreatic juices.

It is known that adrenalin stimulates the sympathetic autonomic system, that pilocarpin stimulates and atropin paralyzes the vagal autonomic system. It has been found by several investigators that the glands of internal secretion exercise either an inhibitory or a stimulating effect upon the two nervous systems. The suprarenals, the thyroid, and the hypophysis increase the excitability of the sympathetic nerves, whereas the ovaries and the pancreas depress the sympathetic and stimulate the vagus nerves.

In a general way, it may be stated that the vagal autonomic nervous system predominates, in health as well as in disease. According to Gautrelet, a condition of sympathetic insufficiency exists in tuberculosis, tabes dorsalis and Addison's disease. He reports observing the reverse only in a single case of pachymeningitis.

Utilizing adrenalin and pilocarpin as diagnostic agents, it is possible, through the effect of these drugs upon the sympathetic and vagal autonomic nervous system respectively, to judge accurately the state of visceral tonus in a number of diseases.

VAGOTONIA AND SYMPATHICOTONIA

Hypertonicity and Hypotonicity of the Vagus and the Sympathetic Nervous System.—The vagotonia and sympathicotonia of Eppinger and Hess rest upon a theoretical rather than upon a practical basis, and the establishment of the physiological and pathological antagonism of these two conditions involves so many difficulties in practice that the intricate action of the nerves upon various structures, such as muscles, blood-vessels, glands and organs in general, still remains more or less doubtful and debatable. Hemmeter, in a recent important contribution to this complicated subject, has ably shown that the increased excitability of various human beings to epinephrin, atropin, pilocarpin, ergotoxin, etc., really indicates that the entire vegetative nervous system—the sympathetic as well as the vagal autonomic—is in a state of morbidly increased irritability.

Vagotonia.—Undoubtedly the conception of vagotonia, as emphasized

by Higier, must be expanded, for there are many exceptions to the rule and many cases which do not fit into the pharmacological schedule of the Vienna school. The electivity of certain hormones—meaning products of internal endocrinic secretion—must be restricted, with all due acknowledgment of the significance of these hormones for the development of somatic neuroses. Numerous mixed and transitional forms may be observed between the two groups of sympathicotonia and vagotonia, as formulated by Eppinger and Hess, who interpret the latter as a functional disease of the vagal autonomic system. From the clinical viewpoint, vagotonia is a permanent state of tonic excitability of the vagal autonomic system through which the affected organs are maintained in a condition approaching a state of autonomic irritation. The symptoms develop upon the basis of a vagotonic disposition, usually met with in youthful or middle-aged individuals who are extremely susceptible to injury of the autonomic system and apt to suffer from more or less pronounced vagotonic symptoms, such as bronchial asthma, bradycardia, vasomotor angina pectoris, etc. This irritability usually diminishes with advancing years, and the vagotonia improves or disappears entirely.

The genesis of so-called vagotonia is explained by Higier as follows: The state of increased autonomic tonicity is probably caused by some product of the internal secretions. Adrenalin is a well-known and highly active sympathicotonic agent. Thyroidin and infundibulin or hypophysis-extract act partly upon the vagus and partly upon the sympathetic nerve. The pancreatic juice and the cholin of the suprarenal cortex possess purely vagotonic properties, but exert an extremely elective effect, by not acting upon the entire vagal autonomic system, but essentially upon the metabolism (pancreatic juice) and upon the blood-pressure and the pupil (cholin). Higier calls attention to the large amounts of these substances which would be required for the maintenance of permanent tonicity in the autonomic system, and emphasizes the great improbability—in contradistinction to adrenalin—of these substances being entitled to the physiological rôle of a theoretical “autonomin” or “autonomic vagotonin.”

A fundamental contrast undoubtedly exists between the sympathetic system, in the restricted sense of the term, and the remainder of the autonomic system, but the anatomophysiological and the pharmacological system are not entirely identical, the contrast between the sympathetic and the autonomic system not manifesting itself in the same way pharmacologically as physiologically. Instead of finding a uniform expression in all the affected end-organs, the increased irritability of the vegetative nervous system tends to manifest itself in a great variety of combinations.

Vagotonia is considered by Eppinger and Hess to be the expression of an inferior constitutional make-up. They emphasize its frequent coincidence with states undoubtedly due to constitutional inferiority of the organism.

The *physiological vagotonia of sleep* is ingeniously interpreted by

Higher as due to the fact that the striated muscular system is at rest during the night, and its blood supply benefits the unstriated muscular system, with the result that the latter functionates more actively during sleep. As a matter of fact, sleep is characterized by a certain hyperirritability of the autonomic nervous system, illustrated by narrowing of the pupils, slowing of the pulse, a tendency to perspiration, asthmatic and colicky attacks, onset of labor pains, and other phenomena pointing in the same direction.

Sympathicotonia.—The sympathetic or vasomotor constitution, which is normal or practically normal in women, is also found in a fairly large number of men, where it constitutes a stigma of femininism. It is characterized by a certain number of anatomical and, especially, physiological anomalies, chiefly of the nervous system and the cardiovascular apparatus; these may be constant, or may manifest themselves only under certain conditions. The typical features of this constitution, with a predominance of psychic, cardiovascular, vasomotor and vasosecretory symptoms, clearly show an exaggerated irritability of the entire sympathetic nervous system.

In the production of this so-called "sympathetic type," a very important part is played by *heredity*. Savini is inclined to interpret it in man as a stigma of femininism and therefore of organic degeneration, although the majority of individuals in whom it is met with possess a normal or even superior intelligence.

In these individuals there exists a mutual and reversible influence between the function of the thyroid gland and that of the nervous system, forming a vicious circle; in other words, there is a mutual abnormal reaction. Hyperactivity of the thyroid puts the nervous system into a state of evident hyperirritability which, in its turn, influences the entire organism and keeps the thyroid in a state of overactive secretion. There is reason to believe that similar relations must exist between the other glands of internal secretion and the nervous system. This adds greatly to the complications of physiological and, especially, of pathological problems along this line of inquiry.

The most characteristic clinical feature of this constitution is *cardiovascular irritability*, which is never accompanied by progressive cardiac insufficiency. Individuals of the sympathetic type are apt to experience precordial distress and palpitations. Under the influence of emotion or fatigue they experience a severe throbbing, not only in the precordial region, but also in the head. The impetus of the heart apex is plainly visible and of great amplitude, but of moderate or small resistance to the finger. Radioscopic examination shows still more distinctly the liveliness and exaggerated amplitude of the cardiac contractions, the difference between the minimum size (systole) and the maximum size (diastole) of the heart-shadow being much greater than under ordinary conditions. The first sound has a peculiar quality, being sometimes slightly indistinct but as a rule somewhat musical, and it may be accompanied or even replaced, after a fatiguing exercise, by a soft murmur, usually perceptible at the apex, more rarely at the base. The readiness with which

this murmur is established clearly indicates the weak tonus of the myocardium.

The pulsations of the superficial arteries are very evident, especially in the presence of fatigue and emotion. The pulse is somewhat bounding, with a tendency to dicrotism, and the number of pulsations per minute is normal or rather slightly above normal. Almost invariably one finds instability of the pulse, irregular respiration, orthostatic tachycardia, and considerable acceleration during fatigue and emotions. The existence of capillary pulsation is often demonstrable, and the arterial tension—both maximal and minimal—is usually slightly diminished, at any rate keeping to the lowest values of physiological variation.

The *exaggerated vasomotility* manifests itself in different ways. In the first place, there is more or less pronounced, sometimes extremely marked, *dermographism*, also known as *urticaria factitia*, or *graphic urticaria*. This appears in predisposed individuals when the finger nail or a suitable instrument is passed over the skin. Pale or white elevations make their appearance, bordered on each side by a red streak. This condition has also been described as *autographism*, and corresponds to the *raie meningitique* or meningitic streak of French writers. Another indication is furnished by the sudden and powerful, although spontaneous, and transitory *congestion* of the head which suddenly appears in these individuals when under agitation or excitement, as for example, when they become the object of attention. This congestion of the head is always accompanied by a peculiar psychic state of embarrassment and distress. There is sometimes hyperhidrosis of the head and neck. The predilection of the congestion for the head is very characteristic. It is sometimes replaced in certain individuals by sudden anemia, as a paradoxical reaction.

Other organs are equally susceptible to the anomaly on which this constitutional type is based. Individuals of the sympathetic type, especially when under the influence of emotion, often have polyuria, attacks of diarrhea or, more correctly speaking, "colicky reactions."

Individuals of this type appear younger than their actual years, and senile changes are delayed. The vasomotor constitution predisposes, however, to neurasthenia with well-marked functional disturbances (cardiac, digestive, etc.) and to various ties. All neuropaths and psychopaths usually have a very unstable sympathetic nervous system, this sympathetic instability being often very pronounced, even in simple neurasthenia, but reaching its highest degree in the symptomatology of exophthalmic goiter.

In individuals of the sympathetic or vasomotor type, the infectious diseases, even when not severe, give rise to a marked tachycardia. Tuberculosis, especially in patients who at the same time possess a narrow thorax, finds a favorable soil for its development in a grave, rapidly progressive form.

All the typical signs of this type of constitution—notably the psychic, cardiovascular, vasomotor and vasosecretory symptoms—plainly indicate an exaggerated irritability of the entire sympathetic nervous sys-

tem. There is an impairment of the regulatory processes, characterized by the direction of the nervous energy exclusively toward the accelerator nervous system of the heart (sympathetic), and the ready occurrence of a diminished cardiovascular tonus, resulting in tachycardia and cardiac dilatation with functional insufficiency.

Nehl, writing on the *influence of the nervous system on the pigment contents in the skin*, emphasizes the undoubted part played by nervous factors in pigmentary anomalies in the human subject. Disturbances of the vegetative nervous system may lead to pigmentation, for example, in exophthalmic goiter or in pellagra, also in Addison's disease, through a change in the suprarenals and their vegetative innervation. The pigmentations of *sclerodermia* and *facial hemiatrophy* are referable to irritative conditions of the sympathetic fibers in the peripheral nerves; interruption of these fibers, in lesions of the cervical sympathetic, causes unilateral canities; resection of the uppermost cervical sympathetic ganglion may be followed by a loss of pigment in the iris. One occasionally sees a patch of gray hair on the side of the head of patients afflicted with migraine, hemierania and trifacial neuralgia.

The name "*sympathoses*" is applied by Laignel-Lavastine* to generalized manifestations of sympathetic functional disturbances, in contradistinction to local sympathetic syndromes. Sympathoses may be simple or compound, according to whether a single function or several sympathetic functions are impaired. A distinction is made between disturbances of sensation, circulation, unstriped muscle, secretion, and trophic disturbances. Among the last-named disturbances, the author includes the arthropathies and bony fragility of patients suffering from tabes dorsalis or syringomyelia. Raynaud's gangrene is also interpreted by the author as a trophic sympathosis. Among the secretory disturbances, those of the external secretions are easily recognized; disturbances of internal secretion consist in certain types of diabetes, exophthalmic goiter, and Addison's disease.

Anaphylaxis.—Anaphylaxis is a term derived from a Greek word meaning protection, and is applied by medical writers to a great variety of conditions. One form of anaphylaxis is an acquired hypersensitivity to the absorption of undigested protein through the intestinal mucosa, or to a parenteral introduction of albuminous substances which have been previously injected on one or several occasions, so that violent phenomena of reaction and even death may follow. The phenomena of anaphylaxis, as pointed out by Pottenger, are apparently those which express themselves, as far as the body structures are concerned, through the greater vagus division of the vegetative nervous system, in the form of *vagus stimulation*. The relationship which general anaphylaxis bears to the vagus suggests a close affinity between this branch of the nervous system and various clinical phenomena, such as hay-fever, asthma, urticaria, and shellfish-poisoning, as well as to the phenomena which take place during visceral inflammation. Pottenger very properly emphasizes the fact that the vegetative or involuntary

* See Laignel-Lavastine, Les sympathoses. La Presse méd., No. 77, 1913, p. 767.

nervous system is not wholly divorced from central control, as is shown by the location in the brain of many of the centers which control certain actions on the part of internal organs.

Other symptoms of anaphylaxis are erythema, fever, vomiting, leukopenia, shock, and especially angioneurotic edema of the face, hands, feet, back, or even of the larynx. It is noteworthy that anaphylaxis often occurs in the young, in children who are asthmatic or have laryngeal spasms and in those with status thymolympathicus. Vagotonia is common in the young and is often found in asthmatic children. It may be seen in laryngeal spasms and not infrequently accompanies status thymolympathicus. Furthermore, the autonomic system controls the vasodilators, and the skin symptoms are probably the expression of a vagus hyperirritability. All the above symptoms which occur in anaphylaxis are equally significant of vagotonia. Finally, **atropin** and **adrenalin** have both been found useful in anaphylactic shock, and as it is known that the first paralyzes the vagal autonomic nervous system while the second produces practically the same effect by stimulating the sympathetic system, it is reasonable to conclude that this favorable action is due to their influence upon the vagotonia.

The peculiar toxemia manifesting itself in the cutaneous lesions known as urticaria may find its etiological factor in the principle of anaphylaxis. Persons of the neurotic type are especially prone to eruptions of an urticarial character.

VASOMOTOR AND TROPHIC CENTERS

Among the centers hitherto recognized in the spinal cord, we have, as is well known, (a) vasomotor centers, and (b) trophic centers. In late years, the relationship between these and the sympathetic centers has been more definitely established. Observations made during the world war have added valuable data. The influence of the sympathetic system upon the equilibrium normally maintained among the glands of internal secretion, while not definitely fixed, is probably the determining factor in certain pathological states clinically recognized.

Vasomotor Centers.—On clinical grounds, we have reason to believe that there is a controlling vasomotor center, probably in the cerebral cortex. If it is not in the gray mantle, it is somewhere in the motor pathway, not improbably in the area of the central ganglia. This seems the more likely since in purely central lesions of the motor area—cortical or subcortical—we have the vasomotor paralysis restricted to the paretic parts of the body. This condition of vasomotor paresis is not the result of local stasis; not only does it appear very early in many cases, but it is occasionally concomitant with the paralysis.

A case of brain tumor, in a series observed by the author at the Montefiore Hospital, showed this phenomenon of vasomotor paralysis synchronous with the transitory paresis. This vasomotor center has been referred to various parts of the brain—to the caudate nucleus, the

corpora quadrigemina, etc.—but the existence of one center only is doubtful. Cortical vasomotor fibers probably accompany the motor and the sensory tracts down through the internal capsule, thalamus, etc., through the pons and medulla, and it is not unlikely that centers are situated at various levels. The ganglion cell groups, as we know, lie in the middle of the gray substance of the cord and through the rami communicantes, and passing through the spinal roots, connect the associative fibers with the sympathetic. From the cord doubtless pass vasomotor and vasoconstrictor fibers. We have not yet definitely learned the variety of activities in the interplay of these two centers, nor are we able to utilize the vasomotor phenomena to any extent for localization diagnosis.

In many cases the vasomotor disturbances are peculiarly transitory. In a case of tumor at the base of the brain, recently observed by the author at the Montefiore Hospital, in which there was marked functional involvement of the hypophysis, the evanescence of the vasomotor symptoms was most striking; there was at times a suffused blush with surface rise of temperature in one or both cheeks, again a suffused duskiness; at other times there was a marked cyanosis of one or of both lower extremities, particularly from a point somewhat above the knee. In this case, variable dryness and scaliness, together with a peculiar induration, suggested vasomotor and trophic involvement.

The association of these symptoms with the sympathetic system and the relationship of the latter to the glands of internal secretion—especially the hypophysis—are among the vital problems of neurology. The connection between the vasomotor and the trophic center, their central localization, the influence of these centers upon certain tissues—especially articular and cutaneous—are suggested by such diseases as scleroderma, spondylitis and arthritis deformans, as well as by some other arthropathies. Furthermore, the presence of trophic and vasomotor symptoms in such central lesions as tabes dorsalis, syringomyelia and myelitis indicates centers for trophism and vasomotility.

Trophic Centers.—True trophic centers may be assumed at various levels of the cord. In tabes and in syringomyelia the nutritional modifications of the bones, nails, hair and skin are determined entirely by the changes in the central nervous system. Whether this is through the sympathetic or vagal autonomic system and whether through the motor or sensory nerves, the essential control is through the ganglion cells of the cord. As in the case of vasomotor influence, no matter to what extent relay systems from the cortex to the periphery play a rôle, the essential fact obtains as stated. The various manifestations of scleroderma, for example, point unequivocally to the nervous system as the seat of this disease. The internal secretions in their interplay may assert their influence. Nevertheless the foundation of the disease upon true structural change within the central nervous system, although as yet undemonstrable, seems to be almost established when we compare the various peripheral changes in scleroderma with similar clinical phenomena in diseases of undoubted nervous origin. The indirect influence of endocrinic balance is certainly obtrusive, for the cutaneous variations in

the true tropho-edemas, in myxedema, the anomalies of adipose deposits in dystrophia adiposogenitalis, in general symmetrical lipomatosis, etc., indicate primarily the influence of the nervous system and, secondarily, anomalies of the internal secretions. In the cases of dystrophia adiposo genitalis the glandular secretion of the hypophysis seems to be primarily responsible, but nevertheless the nervous elements of the organ are undoubtedly first affected.

An interesting combination of trophic, vasomotor and secretory anomalies is observed in scleroderma. The strictly peripheral changes in the bones, tendons, subcutaneous tissue, giving rise to the picture of sclerodactylia, the ulceration and gangrene as part of the latter picture, find some analogy in syringomyelia; in fact, it would seem as if there must be a process within the cord itself directly giving rise to this picture. The almost complete absence of sensory changes in scleroderma, notwithstanding the extensive trophic changes, contraindicates any underlying peripheral nerve disease.

Secretory Function of the Sweat-glands.—Associated with the vasomotor and trophic centers of the cord—probably also in large part under the influence of the autonomic system—is the secretory function of the sweat-glands. Some writers lay stress upon the probable presence of a cortical center for this function. In certain organic lesions of the brain the corresponding paralyzed member of the body has shown anomalies of sweat secretion. Unilateral hyperhidrosis of the right side of the face was observed in a case of glioma of the frontal portion of the left hemisphere. A comparative study of the observations suggests that the secretory changes were due to nerve influence through the sympathetic system.

SCLERODERMA

Etiology.—Scleroderma is a trophoneurosis, characterized by a change in the normal structure of the skin and of the subcutaneous cellular tissue. It occurs in adult life, more particularly in women; a neuropathic tendency and arthritic constitution seem to favor its development. Psychic causes, prolonged nervous disturbances, exposure to cold, menstrual irregularities, repeated pregnancies, disturbances of the ductless glands and infection have all been considered etiological factors, but can only be regarded as exciting causes.

Symptomatology.—**CLINICAL HISTORY.**—The disease begins as a diffuse swelling of the skin and of the subcutaneous cellular tissue—a sort of hard edema, which may spread over the entire integument. The affected skin has a wood-like consistency and is no longer movable on the underlying tissues. All movements, including respiration, may be more or less seriously impeded. This hypertrophic stage is followed by the atrophic stage in most cases, although occasionally the subsidence of the induration is the first step toward a regression of the disease.

The *onset* of diffuse scleroderma is usually insidious, with painful tingling sensations in the limbs, especially in the hands, and various

vasomotor manifestations, but no actual inflammatory symptoms. There may be a slight rise of temperature, without notable impairment of the general health. The disease proceeds slowly toward a gradual absorption of the tissues, which become thin and atrophic. The distribution of the hardened patches in the hypertrophic stage is always more or less symmetrical, with indefinite outlines. Well-developed sclerodermic lesions may terminate in deformities, deviations and actual mutilations. The face in pronounced cases assumes the appearance of a wax cast; the nose, lips, eyelids, cheeks, ear lobes and tongue are thin, shortened, atrophied and retracted, according to the graphic description of Déjerine. The writer has observed, in early cases, a peculiar parchment-like, characteristic feeling, especially easy to recognize in the midline, about the chin and, by careful palpation, on the upper lids.

Sclerodactylism represents an early and constant localization of scleroderma and was well-marked in the case of scleroderma which is described in detail in this article. Besides the cutaneous sclerosis, there is a diminution in the size of the fingers, with osteo-articular changes of the phalanges, or even partial absorption. The affected fingers are contracted and of a purplish color, due to local asphyxia, which occasionally culminates in gangrene and mutilation. The wrist may become fixed and the elbow maintained in flexion by the sclerotic process, which is sometimes associated with considerable muscular atrophy, with resulting paralysis of the hardened muscles. These features of the disease are well defined in a case now under observation in the Montefiore Hospital.

CASE I.—The patient, an inmate of the Montefiore Hospital, is a young unmarried woman, 28 years old, of Austrian extraction. Her personal and family history are essentially negative. The disease responsible for her present condition began at the age of sixteen years. There were no direct prodromata and no variation of health that might be regarded as preceding the somatic changes. The initial phase—rather abrupt in onset—was a swelling and redness, soon attended by a peculiar subjective feeling of stiffness of the toes; then a tight, or stiffened feeling about the knees was noted, followed by pain. Progressive stiffness of the smaller articulations of the feet and a tenseness of the cutaneous tissues soon so restricted motion in these parts that the patient could hardly flex the limbs, and sitting became difficult and painful.

Coldness of the distal parts of the lower extremities and a feeling of weakness in the legs became pronounced. Involvement of the hands succeeded the earliest symptoms by about three months; the fingers became gradually stiffened and the psychro-esthesia in the hands was attended by an objective coldness of the surface. At frequent intervals there followed paroxysms of digital pallor, with pain and tingling sensations in the fingers. In these attacks the blanching was succeeded by congestion and cyanosis, thus simulating Raynaud's disease, a related trophoneurosis. The hands became entirely useless by reason, especially, of a growing weakness and a rapidly progressive contraction of

the forearm flexors and a tightening with tension of the skin of the hands. So rapid was this process that within a month it had reached an advanced degree. The symptoms all progressed, reaching such severity as to render the patient practically bedridden for a time.

Facies: Two years after the onset the soft tissues about the oral opening and jaws became changed; the tension, due to trophic changes of the skin, restricted the opening of the mouth and the moving of the jaws. Gradually the skin of the entire face and forehead and upper



FIG. 3.—SCLERODERMA IN THE LOWER EXTREMITIES.
Showing osseous changes and diffuse irregular bone atrophy.

part of the neck became thin, tense and hard, having to the touch the smooth, marble-like feeling characteristic of this disease.

Teeth: The teeth had a peculiar luster, especially the front teeth, which showed a bluish transparency, giving them an unnatural tinge. This was particularly true of the frontal and lateral incisors. The mucous membrane, especially of the lips, was dry and tense and had a pale, anemic color.

Movements: The general stiffness of the entire cutaneous mantle so affected the movements, that in attempting to lift the patient one had the impression of raising a wax doll.

Hair: The hair of the head felt dry and wiry to the touch, quite in contrast to the condition the patient described before her illness, when the hair was silky and soft and the scalp well lubricated with its natural secretion.

Lower Extremities: The left dorsalis pedis artery pulsated distinctly. At this particular examination the extremities, including the hands, were warm and only slightly cyanosed, except the soles. The two feet showed a marked contrast, both in appearance and on palpation; the left was fairly white, showing very little evanosis except about the sole of the

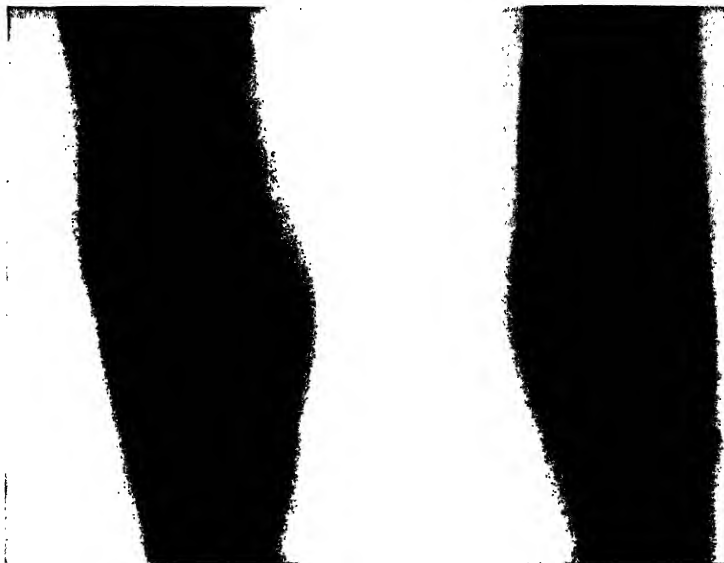


FIG. 4.—SCLERODERMA IN THE LOWER EXTREMITIES.

Showing osseous changes and marked diffuse bony atrophy, associated with fibrous degeneration of peri-arthritic tissues. Slight osseous arthritic changes.

foot, where a marked, diffuse duskiness was apparent, emphasized by the contrast brought out by pressure of the soft parts which, until the blood slowly diffused, left a white, parchment-like appearance. The left foot had a normal warmth, while the right was relatively cold; the duskiness about the sole and heel was much more marked than on the other foot. That the peripheral vascular supply plays, to some degree, an etiological rôle is suggested by the fact that no pulsation of the right dorsalis pedis artery was palpable. However, the fact that there was a thickening and tenseness of the cutaneous tissue, thus making the recognition of the artery by touch somewhat more difficult, must be taken into consideration in determining whether or not the pulsation was altogether absent. The posterior tibial artery was not felt in either foot; it is pos-

sible that the latter vessels could not be felt because of the hidebound condition of the skin. This hidebound condition of the cutaneous surfaces was felt well up into the thighs on both legs.

Abdomen: In the lower abdomen, while the outer layer of the skin had a feeling of tenseness, there was, in contrast to the skin just above it, an appreciable thickness, a distinctly palpable hypertrophy of the cutaneous tissue. The feeling did not suggest material of an edematous



FIGS. 5 and 6.—SCLERODERMA, WITH OSSEOUS TROPHIC CHANGES IN PHALANGES.

character, nor was it like subcutaneous fat, but altogether it suggested the hard surface of thin leather with a somewhat softer composition beneath. The process is probably a true hypertrophy of the subcutaneous tissue, quite in contrast to the atrophy found in other parts of the skin mantle.

Upper Extremities: The fore and upper arms showed very marked brownish pigmentation. There was a pronounced surface growth of hair on the extensor surfaces of the forearms. The brawny induration of the skin, together with the peculiar hypertrophic feeling of the skin beneath, was likewise quite noticeable on the outer surface of the forearms just beneath the elbows; the texture on palpation was very much



FIG. 7.—SCLERODERMA.

Showing teeth which have undergone changes in structure; pigmentation of skin of chest; sclerodactylia with trophic and ulcerative changes of phalanges (osseous and soft parts).



FIG. 8.—SCLERODERMA.

Showing mask face and stone-like fixation; sclerodactylia and trophic changes of hands. The trophic skin and subcutaneous tissue feel like leather to the touch.

like that of an elephant's hide. The hands, on the contrary, showed a diffuse pink luster, while over the strongly flexed and sclerodactylic upper extremities the most pronounced hidebound condition of the skin was manifest. White, anemic skin surfaces, due in large part to impoverished blood supply and pressure of the underlying osseous surfaces, appeared in both hands, and surrounding these were rings of tissue seemingly about to ulcerate. The radial artery was distinctly



FIG. 9.—SCLERODERMA.

Showing parchment-like "cutaneous cuirass"; fringe of gray hair, trophic canities.

felt some two inches above the wrist on both sides, but not at the wrist, doubtless due to the cutaneous thickening. To the touch, the vessel seemed full and normal. The *x-ray* revealed marked changes in the osseous structures. (See Figs. 3, 4, 5, 6.)

Trunk: Brownish pigmentation was almost diffuse over the trunk, being accentuated in symmetrical patches suggesting segmental distribution.

Labia: The labia were hard, indurated to the touch.

Hips: Owing to the ankylosis at the hip joints, the legs could be

separated only about an inch; further local examination, therefore, was impossible. For a while thyroid therapy had a markedly favorable influence upon the symptoms generally. (Figs. 7 and 8.)

CASE II.—Another illustrative case at the Montefiore Hospital was that of a woman 39 years of age, whose symptoms began twelve years previously, following a local trauma. The patient's index finger of the left hand became infected by the prick of a sewing needle. Characteristic cutaneous and ulcerative changes in the fingers dated from that time, and after a few years similar changes appeared in the toes. Then myxedematous infiltration of the skin of the legs and arms gradually developed, the latter being more seriously affected. The indurated



FIG. 10.—SCLERODERMA

Trophic changes with transformation of cutaneous parts to parchment-like tissue.

condition of the cutaneous tissue in the distal portions of the upper and lower extremities produced, on palpation, the typical hidebound sensation of scleroderma. The skin on the trunk likewise began to give to the patient a subjective feeling of tenseness. Trophic changes in the skin and nails of fingers and toes presented characteristic lesions, and brown pigmentary deposits were noted on the abdomen. The vasomotor changes were at times so marked as to suggest the phenomena of an atypical Raynaud's disease. It is of interest to note that subcutaneous injections of adrenalin intensified the clinical symptoms.

CASE III.—Fig. 9 is the photograph of a patient, an elderly man, suffering from scleroderma. The entire trunk, face and extremities were set in a parchment-like cutaneous cuirass. The skin surface of the

upper part of the back was pigmented. A rather peculiar trophic phenomenon observed was a circular fringe of gray hair around a dark central tuft.

Figs. 10 and 11 show marked trophic changes of the lower extremities in another case of scleroderma under observation at the Montefiore Hospital.



FIG. 11.—SCLERODERMA.

Same case as Fig. 10 (more advanced). Note changes in the hands and fingers.

Association with Other Diseases.—Scleroderma may become associated with progressive facial hemiatrophy, and it has also been observed in combination with Raynaud's disease, erythromelalgia and other trophoneuroses. Its coexistence with exophthalmic goiter as well as with Addison's disease has been repeatedly noted. Anomalies of pigmentation, irregular heart-action, albuminuria, attacks of diarrhea, etc., point in certain cases to visceral localizations of the sclerotic process; the influence of the autonomic system is here suggested. The autopsy of scleroderma patients, who usually die from cachexia or as the result

of some intercurrent disease, often reveals changes in the ductless glands—especially in the thyroid and the suprarenals—suggesting that generalized progressive scleroderma may be due to simultaneous functional disturbances of several internally secreting glands (Déjerine). Available evidence still fails to enable one to make definite statements as to which gland is most involved. For example, in one case at the Montefiore Hospital—the one above described as having followed local infection—the subcutaneous administration of adrenalin markedly aggravated the clinical picture.

A case of exophthalmic goiter with patches of scleroderma, in a girl of twenty years, reported by Marinesco and Goldstein in 1913, is of special interest inasmuch as the scleroderma appeared after x-ray treatment of the goiter, together with the onset of trophic disturbances in the form of pigmentation. The undoubted dependence of certain cases of scleroderma upon functional disturbances of the thyroid gland is possibly due to a sympathicotonic action of the thyroid secretion. Cases of scleroderma have been placed on record in which marked improvement followed thyroid opotherapy. It would be possible, however, to quote from the literature illustrative cases in support of each of the pathogenetic theories which have been propounded—trophoneurotic, sympathetic, vascular, thyroid, hypophyseal, pluriglandular, etc. The disease is accordingly of rather obscure origin. The assumption of changes of the sympathetic nerve as the cause of scleroderma (Brisaud) or of gross organic lesions in the central and peripheral nervous system (Bruns) is not accepted by Cassirer and Curschmann, who interpret this affection as being probably merely a functional disease of the vasomotor and trophic tracts and centers, not yet entirely understood. The author is strongly inclined to agree with the last mentioned writers.

Traumatism may play a part in the development of a certain number of cases of circumscribed scleroderma, and as generalized scleroderma sometimes follows closely upon a traumatism, a causative relation is very probable. This relation is less evident in the progressive sclerodermas, where the traumatism constitutes only one of the factors of an undoubtedly complex and highly elusive etiology. Here it intervenes, probably, merely as a determining cause, activating one or several pathogenic conditions already present, the influence possibly varying according to the clinical form of the cutaneous sclerosis.

The same condition has been described under the names of scleroderma, scleroma, cutaneous sclerostenosis, cutis tensa chronica, elephantiasis sclerosa, Addison's keloid, cicatrizing scleroma of the skin, Alibert's scleroma, adult scleroma, etc.

A case which has been under the author's observation for several years combines many interesting phases: a generalized scleroderma with sclerodactylia, atrophic changes in the mucous membranes, and almost generalized pigmentation of the skin. This case shows the presence simultaneously of three phases of cutaneous changes.

In cases of scleroderma generally, and especially in one of the cases

which the author is reporting, anomalies of sweat secretion appear early in the disease. The usual hypohidrosis in scleroderma might be regarded as due to local changes in the skin. However, as illustrated by the case mentioned above, the paradoxical phenomenon is sometimes observed of atrophic cutaneous tissue with hyperhidrosis.

Treatment.—As far as a cure is concerned, the therapy now at our disposal has but little to offer. In the light of the modern conception of some observers that the condition is due to glandular dysfunction, one is justified in applying therapy experimentally along these lines. The most promising results have been obtained from the administration of **thyroid extract**, and although reports are variable and indefinite, this therapeutic measure should be tried. The effect of **supra-renal extract** should also be tested. In using glandular extracts under any conditions, one must be guided by the complex of symptoms which in conjunction suggest the nature of the dysfunction, or rather the hyper- or hypofunction of one or more glands. In this field of therapy, there is still a wide difference of opinion as to dosage and as to the particular gland-extract to be given. Individual observation and experience must be our guide. In general, however, the treatment of scleroderma consists in **maintaining the general health** of the patient, and here the familiar **tonics**, especially **arsenic**, **sodium salicylate** and **quinin** are beneficial. When the function of the secretory glands of the skin is deficient, **pilocarpin**, cautiously administered with tonics, is helpful. Local treatment such as **baking**, **mild friction** and **massage with bland ointments**, is ameliorating. General and local **galvanization** is probably of the same value as mild stimulation and is to be recommended as symptomatic therapy. The author has not definitely noted improvement due to any therapy, although amelioration of symptoms has been frequently observed, independent of treatment.

RAYNAUD'S DISEASE

Definition.—Raynaud's disease, a condition also known as symmetrical asphyxia or gangrene, is a *vasomotor* or *trophic neurosis*, in which cyanosis, local asphyxia, and gangrene of the extremities, often symmetrical, appear and are associated with neuralgic pains, dysesthesias, diminished sensibility, a subjective sensation of cold, etc. Under the same heading are also included those cases in which obliteration of the peripheral blood-vessels occurs as the result of intoxication (ergotism) or of anatomical vascular changes, such as arteriosclerosis or thrombosis.

Etiology.—The disease shows a predilection for women between 18 and 40 years of age. It has been observed in neuropathic, emotionally unstable, hysterical and alcoholic individuals, in certain types of mental disease, in epilepsy, in the course of pulmonary tuberculosis, leprosy, leukemia, syphilis, and diabetes. Although in many cases no etiology is demonstrable, the onset and occurrence are undoubtedly influenced

by exposure to cold, menstrual anomalies, and emotional disturbances.

The pathogenesis of symmetrical gangrene of the extremities is still a matter of controversy. Although the majority of authors are inclined to refer it to a process within the nervous system—central, peripheral, or especially *vasomotor*—others suggest that lesions of the arterial system may furnish the causative factor. The observer who gave his name to the disease described it as a form of dry gangrene, characterized by the double fact that it is independent of all demonstrable anatomical change of the vascular system and that it invariably affects symmetrical parts. Since that time (1862) a number of clinical and anatomico-pathological observations have been published of cases in which the vascular system was distinctly altered, supporting the arterial theory of symmetrical peripheral gangrene.

The pathogenesis of local asphyxia was referred by Raynaud to a vasomotor spasm, inducing transitory ischemia, an excessive duration of this angiospasm being followed by the onset of gangrene. According to the present state of our knowledge, angiospasm alone can never lead to gangrene and can only be regarded as a secondary factor in its production.

On the other hand, arterial lesions alone are capable of causing gangrene, and it is not impossible that the transitory angiospasm which produces local syncope and asphyxia in cases which do not terminate in gangrene is itself the result of a very minute latent arterial lesion. As the vasomotor nerves of the limbs follow the walls of the arteries especially, it may be that an arterial lesion excites and localizes a temporary or permanent angiospasm either directly or reflexly. It is difficult to determine the relations existing between the character or extent of the arterial lesion and the duration or severity of the spasm.

In the opinion of some observers (Vulpian) vascular spasm does not absolutely require the intervention of vasomotor centers for its production, but may be caused by ganglia situated on the vasomotor fibers which accompany the blood-vessels. Other writers have referred the disease to neuritic changes of the collateral nerves of the fingers. Buerger, who does not accept the vascular origin of Raynaud's disease, states that in this and allied diseases the vasomotor and trophic disturbances are the outcome of irritative and exhaustive processes of the sympathetic nervous system.

Symptomatology.—The disease is characterized by a symmetrical arrest of the capillary circulation in the extremities, especially in the fingers, sometimes terminating in dry gangrene. The mildest stage is that of so-called local syncope, in which one finger of the hand becomes pale and suddenly turns cold, either without any apparent cause or on simple exposure to the air. The skin becomes an opaque or yellowish white; the surface temperature is diminished; cutaneous sensibility disappears, and the affected finger remains, for some minutes or even for several hours, as if paralyzed or "dead." The condition is entirely painless. Gradually the circulation is reëstablished, to-

gether with a return of the normal color, warmth, and sensibility. In a more advanced stage, the finger presents a bluish or purplish discoloration, pressure on which leaves a lighter spot, persisting for a certain length of time. Severe tingling or burning pains are usually present in these cases. The return to the normal condition is accompanied by intolerable prickling sensations, and the skin turns a bright red before the normal color is restored. The two stages may be associated, alternating in the same localities, or existing simultaneously in the same finger, in the form of white and purplish spots. In the interval between the attacks, which as a rule affect both hands symmetrically in one or several fingers, the tissues present no demonstrable changes.

In peripheral gangrene—the highest degree of Raynaud's disease—the extremities pass from a pale to a bluish or dusky red color, and the purplish discoloration of the finger tips is often visible through the nails. Tingling and painful sensations occur in extremely violent paroxysms, during which the discoloration deepens. The affected parts are very cold, the local temperature being diminished by several degrees, while directly above the wrist and the palm of the hand are rather warmer than normal. At the end of a few days the fingers become almost black and the discolored veins are seen standing out prominently along the affected extremity. Well-marked trophic disturbances make their appearance, in the form of small isolated or confluent vesicles and sloughs. Sometimes these vesicles rupture, exposing the reddened dermis; in other cases they dry up and are shed, leaving superficial ulcerations which gradually heal, the finger again becoming warm and resuming its normal color until the next attack. The skin may assume a horny or parchment-like appearance and condition, so that the finger finally resembles that of a mummy. The whole process suggests the various phases of hyperemia and ischemia, through which the parts pass when exposed to the prolonged action of cold, as in freezing. After a series of attacks, the fingers are seen to be very thin, hard, and tapering; their extremities are covered with small white cicatrices, and the nails present trophic disturbances. The majority of the cases terminate in recovery.

Less common localizations of Raynaud's disease than the fingers and toes are the nose, the ears, the cheek bones, the heels, the external malleoli, and the coccyx.

CASE.—A typical case of Raynaud's disease which the author observed in Lassar's clinic in Berlin and reported in 1896, was that of a neuropathic woman 37 years of age, who showed an advanced, practically symmetrical vasomotor disturbance, manifesting itself in the peripheral ends of the four extremities, combined with acroparesthesia and severe pains, recurring periodically at certain seasons of the year. The course of the attacks was characteristic of the disease, beginning with dysesthesia, pallor, and progressive pain; the pallor gradually passed into cyanosis, while the pains became replaced by a "dead"

sensation. The patient's first severe attack appeared as a sequel of severe emotional disturbances, illustrating the well-known connection between psychic conditions and vasomotor excitability.

Diagnosis.—A sharp distinction is made by Buerger between Raynaud's disease and *thrombo-angiitis obliterans*, a condition which may be associated with clinical manifestations closely resembling the former, but depending upon vascular occlusion. There are cases of thrombo-angiitis obliterans in which vasomotor phenomena preponderate, and there are other cases which are associated with trophic disturbances exclusively. The prominent features of thrombo-angiitis obliterans are summed up by him as the apparent dependency of the vasomotor symptoms upon variations in temperature, the chronicity of the manifestations, the absence of pain in some of the cases, and the absence of the paroxysmal attacks so characteristic in Raynaud's disease. Unlike the latter, thrombo-angiitis obliterans is not a vasomotor neurosis.

Symptoms of acute Raynaud's disease associated with paralysis of the extremities were observed by Alessandri and Mingazzini in a considerable number of cases of victims of a recent Italian earthquake. These cases always occurred under the same conditions, doubtless attended by great psychic trauma, in persons who had been buried for two days or longer under the ruins. Circulatory disturbances were present, due to the crushing sustained by the extremities, together with exposure to damp and cold.

Clinical Varieties.—A distinction can be made between two degrees of Raynaud's disease, according to the severity of the manifestations: (a) simple asphyxiation of the extremities; (b) peripheral gangrene. The former may supervene in connection with neuroses, such as hysteria or epilepsy, but it has also been observed in the course of various organic nervous diseases, and at the onset of scleroderma. It is usually regarded as a vasomotor neurosis, however, because the majority of cases are free from all other nervous manifestations.

Treatment.—Constructive treatment along general constitutional lines is indicated. The patients are, as a rule, of neuropathic makeup and require **regulation of their habits and manner of living**. Recognition of the probable causative factor underlying the condition will often suggest direct therapy. In general, a **generous but regulated diet** and attention to **hygienic conditions**, together with therapy designed to influence the peripheral circulation, especially **amyl nitrite** and **nitroglycerin**, are indicated. In the early stages the **application of friction** is advisable; later the treatment may become surgical. **Electricity**, in the form of high frequency, is valuable in some cases; the **galvanic current** can be utilized by placing the affected parts in hot water, in which the cathode is immersed. The anode is placed at an indifferent point, and a strong current is allowed to pass for about ten minutes. In this way the influence of the cathode as a vessel dilator may be of service in establishing circulation.

Recently L. Veillet reported a form of **surgical therapy**, consisting

of cutting the peri-arterial nerves of the sympathetic system. The sympathetic nerves were "peeled" from the right brachial artery. Veillet reports recovery from well-defined symptoms in both upper extremities; symptoms in the lower extremities were naturally not influenced. The observer regards the improvement in both hands, although the operation was unilateral, to be explained by the physiology of the sympathetic function in its influence upon symmetrical structure. The procedure appears to be radical, but taking into consideration the great disturbance in function and in subjective feeling caused by symmetrical gangrene, any form of therapy that offers relief is warranted.

FACIAL HEMIATROPHY

Definition.—Facial hemiatrophy (*hemiatrophia facialis progressiva*) is a term which should be applied only to cases in which all the tissues of one side of the face, the osseous parts included, are affected by a slowly progressive atrophy. The wasting may primarily affect a limited area or be confined for a time to one tissue.

Facial hemiatrophy is variably interpreted as a trophoneurosis, originating in the sympathetic nervous system, or as a primary disturbance of the fifth nerve, beginning in the gasserian ganglion. Certain autopsy findings suggest that the disease is sometimes caused by neuritis of the fifth nerve with special involvement of the descending intracranial roots.

Etiology.—This very uncommon disease usually begins at an early age, often in the form of atrophic patches which gradually extend. Several authors have compared it with scleroderma. Cases have been reported in which facial trophoneurosis coincided with scleroderma in extensive patches.

Traumatism of the face and skull represents the best known etiological factor, although preceding infectious diseases are repeatedly mentioned. The patients are, as a rule, youthful individuals, for the most part between ten and twenty years of age, the disease being of exceptional occurrence after the age of thirty. The literature contains approximately 150 cases.

The cause of facial hemiatrophy, as shown by a certain number of cases, may consist in lesions of the central sympathetic tracts, which are situated in the periependymal gray matter of the sylvian aqueduct and in the fourth ventricle. This assumption does not exclude the possibility of a facial hemiatrophy being due to an affection of the superior cervical ganglion with its afferent and efferent tracts. Such cases represent a peripheral localization in the same system. The central or peripheral lesions, respectively, probably give rise to differences in the clinical picture. Vasomotor disturbances and inequality of blood supply on the two sides of the face are almost invariably present in cases with a peripheral localization in the superior cervical ganglion.

Symptomatology.—This disease, also known as facial trophoneurosis

or laminar aplasia, consists of a well-marked thinning—without sclerosis or adhesions—of the skin of one-half of the face. The atrophy extends to the corresponding half of the palate, the palatine velum, and sometimes to the tongue. The prominences of the bony framework are likewise reduced. The affected side appears older and is displaced backward. The skin is white or pigmented. The sensibility is intact, but anhidrosis and alopecia are usually present.

CASE I.—A case observed at the Vanderbilt Clinic was that of a young girl of about fifteen years of age who suffered from a facial hemiatrophy with hemiatrophy of the tongue on the same side. There were no objective or subjective sensory changes, nor can the author recall any etiological factors. The asymmetry was observed in the early years of life.

CASE II.—The youngest case in my experience was that of a child three years of age, recently observed through the courtesy of Dr. I. S. Wechsler, of New York. The condition was congenital; there was narrowing of the palpebral fissure with enophthalmos, these abnormalities affecting the left side. This case suggests a lesion of the cervical sympathetic system. There was no history of a birth trauma, although the asymmetries were observed in the early months of the child's life.

CASE III.—An illustrative observation on a case of facial hemiatrophy at an unusually advanced age, in a woman of 41 years, with a history of traumatism, was reported by Neustadter, in 1914. The Wassermann reaction of the blood was negative. Physical examination showed marked hemiatrophy of the entire right side of the face embracing the skin, muscles and bony structures. "The temporal muscle seems to be completely gone. The skin is rather thin as compared with the left side of the face and of a yellowish-brown hue. The facial muscles and those of mastication are considerably involved and, to some extent, also the right half of the tongue. The muscle tonus of the affected group is good and its functions are not impaired. The electrical reactions to the faradic and galvanic currents are normal. The bones on the affected side, as shown by the x-ray, are decidedly smaller, the inferior maxilla, the right orbit and the right frontal sinus being nearly half the size of those on the left side, giving the face a peculiar asymmetry."

CASE IV.—A very instructive case of facial hemiatrophy, with contralateral Argyll Robertson pupil, in a boy of 11 years, with a normal heredity and a negative Wassermann reaction, was reported by Langelaan (1913). There were well-marked hemiatrophy of the left side of the face, especially in the region of the upper jaw, considerable mydriasis, reflex rigidity of the pupils, slight nystagmus, slight insufficiency of the external recti muscles. The observer assumed a lesion at the level of the red nucleus, at the point where the oculomotor nuclei come in contact with the periependymal gray matter of the sylvian aqueduct.

CASE V.—A case of progressive facial hemiatrophy, reported by Krueger in 1916, was associated with sensory disturbances and homolateral tonic-clonic spasms of the masseter muscle. The irritative motor phenomena are most readily accounted for by a lesion of the trigeminal nucleus.

Treatment.—Treatment is, as a rule, productive of no direct results. **Sympathectomy** is apparently of no value in influencing the condition. **Subcutaneous injections of paraffin** are recommended for cosmetic effects but require care and skilled technic. Embolism of the central retinal artery has been repeatedly reported as a result of this procedure. **Injections of oil and vaseline** have likewise given good cosmetic results in selected cases. The **galvanic current** is claimed by some to have a beneficial effect. The author is skeptical of any benefit to be obtained from electrotherapy and would urge great reserve in performing operations for cosmetic purposes.

ERYTHROMELALGIA

Definition.—Erythromelalgia is an angioneurosis characterized by attacks of pain, redness and swelling of the distal parts of the limbs or of the toes, respectively. It is a symptom noted in various central and peripheral nervous diseases. The name is derived from two Greek words, meaning *red* and *limb*. Weir Mitchell was the first to describe this condition (1872):

Akromelalgia is a *vasomotor neurosis* probably identical with erythromelalgia and consisting in redness, swelling and pain in the fingers and toes, combined with headache and vomiting.

Etiology.—The pathogenesis of erythromelalgia still remains rather obscure. The somewhat uncertain etiology seems to be referable especially to diseases of the nervous system and to vascular affections.

Physiology teaches that the vasomotor function is governed by the sympathetic and the cerebrospinal systems, the vasomotor reflexes and especially the thermic peripheral reflex being controlled by various centers, among which the bulbomedullary centers exert a predominant influence.

Various *pathogenic theories* of erythromelalgia have been propounded, such as the encephalic, medullary, vascular, and neuritic theories. Among these theories, the one most in conformity with the different anatomical, physiological, clinical, and anatomicopathological findings is the medullary theory, although there is a remarkable constancy of the vascular lesions. On eclectic grounds, the following theory suggests itself:

Erythromelalgia and Raynaud's disease are two affections apparently related to the same disturbances of the thermic peripheral vasomotor reflex. They are caused by an alteration of the medullary vasomotor centers. These centers, which represent the medullary origins of the

sympathetic system within the spinal cord, are localized in the basal region of the medullary horns, and especially at the level of the intermediate lateral tract of Clarke. The medullary or cord lesions in erythromelalgia may be secondary to vascular changes.

Symptomatology.—The central nervous system is free from gross changes in idiopathic cases of this extremely uncommon vasomotor neurosis, which must be referred, according to Cassirer, to irritative conditions in certain sensory vasomotor and secretory tracts or centers. These patients are apt to suffer at the same time from more or less characteristic symptoms of hysteria. The clinical picture shows progressive acute attacks of severe pain, followed sooner or later by circumscribed redness and tumefaction of the ends of the extremities. As a rule, residues are left by each attack until the condition finally becomes definitely established and permanent.

The disease is characterized by painful attacks or paroxysms, generally localized in the extremities, and accompanied by local congestion, which is indicated by redness, swelling and a local rise of temperature. Both sexes are about equally susceptible to erythromelalgia, which is most frequent between the age of thirty and forty years.

Prognosis.—Erythromelalgia is an essentially chronic and extremely distressing condition of indefinite duration. The vital prognosis is dependent upon that of the organic diseases sometimes associated with it, which often lead to death.

Pathology.—Anatomicopathological investigations show the constant presence of vascular lesions, in the form of arteriosclerosis and endarteritis. In some cases, degenerative changes in a few peripheral nerves have been demonstrated, perhaps secondary to the vascular lesions. A very small number of autopsies have shown medullary lesions. Auerbach (1897) found a degeneration of the spinal roots of the lumbosacral nerves. Lannois and Porot (1903) observed multiple foci of cerebral softening and atrophic lesions of the gray matter of the posterior horn of the cord.

Erythromelalgia and Raynaud's disease are two vasomotor affections with absolutely opposite symptoms but with an analogous etiology, clinical associations and pathological anatomy. The two diseases often co-exist, and seem to represent an exaggeration of the phenomena of congestion and ischemia.

THERMALGIA (CAUSALGIA)

Definition.—Causalgia, or thermalgia, is a definite peripheral nerve syndrome of neuralgic character attended by violent pains of a burning nature, as though due to thermic stimuli.

Etiology.—The condition is practically limited to injuries and irritation of the median and, less often, of the sciatic nerves, and is a possible result of partial division of the nerve and excessive formation of scar tissue. A number of cases have been added by the World War

to the pioneer reports of Weir Mitchell, from observations made during the American Civil War.

Thermalgia was discussed in all its bearings in a Report to the Medical Research Committee by John S. Stopford in England. This author emphasizes the constant presence of partial division of the nerve and intraneural fibrosis in these cases. The restriction of thermalgia to injuries of the median nerve or internal popliteal fibers is possibly due to the larger number of vasomotor fibers in these as compared with other peripheral nerves. The character of the pain itself points to a disturbance of vasomotor control, which is likewise suggested by the increased surface temperature, often with cutaneous hyperemia. The explanation of the evident vasodilatation which is present meets with difficulties and must still remain a subject of further investigation. In another contribution, the same observer refers the ultimate cause of traumatic trophoneurotic disturbances to the irritative effect of peripheral nerve lesions on the walls of the adjacent blood-vessels, resulting in defective blood supply to the affected region through the reduction in the caliber of its nutrient vessels.

Symptomatology.—As Tinel states, the causalgic syndrome almost always accompanies a slight lesion of the nerve, paralysis is seldom found, and the sensory disturbances are absent or slight. The symptoms appear quite suddenly, with burning pain as the dominant manifestation. There are but few trophic disturbances. The pain appears early, usually just after the injury occurs and gradually increases, reaching a maximum intensity in two or three weeks. The reports by Tinel, who studied many cases in the World War, are instructive. Patients suffer very intense, persistent pain, which is constant and may occur both by day and by night. As a result the patients often show general weakness and great irritability. Although the injury may be in the hand, wrist or forearm, the pains usually spread to the upper arm, but are especially severe in the hand. External stimuli, such as cold, heat, or even tactile contact, may give rise to very severe pain, and even simple movement of the part may cause pain. That fear and the emotions play a rôle is evident from the fact that these may excite a paroxysm of pain. The general psychic state of the patient may be profoundly affected, probably by the severe pain, and by the many causes that may induce the paroxysm. Intense hyperesthesia is conspicuous. It is a noteworthy fact that while a simple touch or slight pressure of the affected extremity may cause pain, firm compression of the soft parts, either integument or muscles or both, is only slightly or not at all painful. Surface stimuli are painful, deep pressure is not.

The trophic and vasomotor symptoms are not marked. The skin, as a rule, is thin, rather than thickened as is the case in neuritis, and appears white or glossy. It may, however, be red. Tinel describes trophic changes of a characteristic nature. He has observed that the nails, in causalgia of the median nerve, are curved as in neuritis, but are thin and smooth and not thickened; they grow rapidly, and a small, cutaneous, painful swelling forms behind the pulp. Wasting or atrophy

of the peripheral ends of the index and middle fingers may occur. Tinel has observed peculiar trophic lesions such as small subungual ecchymoses and small cutaneous phlyctenæ. Some other rare trophic forms are reported as having been observed during the recent war.

Treatment.—Thermalgia affecting the median nerve is not easily amenable to treatment. The pain may continue for from eight to fifteen months. **Roentgenotherapy** applied to the nerve-trunk or to the roots of the plexus often gives relief, but not permanently. Tinel very properly proposes the treatment suggested by Leriche, namely **denudation of the brachial artery and resection of the sympathetic plexus surrounding the vessel**. Tinel reports favorable results in several otherwise refractory cases. The treatment seems logical if we accept the observer's deductions from his observations in cases of wounds at the wrist and forearm. In these he has found disturbances throughout the area of the cervical sympathetic system, with narrowing and vasoconstriction of the brachial artery. There was also some numbness of the superficial parts on the affected side; hypohydrosis, vasodilator or constrictor symptoms were present, strongly suggesting the presence of a reflex stimulus of the cervical sympathetic system. The **application of moist warmth** to the parts affected by causalgia gives relief.

In causalgia of the lower extremity **arterial denudation of the femoral and resection of the sympathetic nerve plexus** are proposed.

ACROPARESTHESIA

Definition.—A disturbance of the vasomotor innervation, in the form of a vasoconstrictor neurosis, is responsible for the onset of intermittent spasms in the peripheral blood-vessels, especially in those of the hands and feet, more rarely in those of the ears or the nose. The affected parts become pale, cyanotic, or reddened. Various sensory phenomena are associated with this condition. According to the most authoritative statements, these sensory phenomena are always referable to a vascular spasm in the peripheral arteries. A peculiar instability and irritability of the peripheral vasoconstrictor nerves is characteristic of acroparesthesia, usually as a partial manifestation of a general neuropathic diathesis. Curschmann goes so far as to assume the existence of permanent tonic changes in the peripheral arteries in these cases, interpreting the acroparesthetic attacks as the paroxysmal increase of this vasoconstrictor disposition. The opposite tendency, namely a vasodilator disposition, is illustrated by manifestations such as dermographism or "psychic" erythema, due to causes of an emotional nature.

It seems as though a more properly descriptive term for this condition would be *acrodysesthesia*, rather than *acroparesthesia*.

Etiology.—The etiological factors are represented by exposure to cold, arteriosclerosis, alcoholism, and sometimes by local factors affecting the hands and feet (occupational acroparesthesia).

The majority of patients suffering from this vasomotor neurosis are

women between thirty and sixty years of age. A connection with pregnancy, the puerperium or climacteric can sometimes be established, and the disease has been known to follow upon oöphorectomy and hysterectomy.

Trophic disturbances of the skin, nails, and hair, as well as *amyotrophia*, sometimes also tendinous, muscular and articular contractions, or trophic lesions of the bony structure (osteoporosis) are described by French observers in some cases of acrocontracture and acroparalysis due to war wounds. In their opinion, these cutaneous disorders of trophic type are, to a great extent, referable to immobilization of the wounded limb, for they were observed to improve and rapidly subside with the return of motility.*

Symptomatology.—As implied by its name, acroparesthesia seems to be limited to the local sensory and vascular nerve terminals and ganglia. The principal feature of this vasomotor trophic neurosis consists in the gradual onset of dysesthesia of one or several extremities, the patient complaining of a sensation of tingling or pricking in the fingers, more particularly in the finger-tips. The disturbance varies from mere discomfort to well-marked pain and actual interference with work and sleep. The troublesome symptoms may extend from the fingers to the hand, occasionally involving the arm itself. Although the condition is bilateral in the majority of cases, one hand is usually more severely affected than the other, and the fingers are involved to a variable degree, sometimes without the hand being affected. In certain rare cases, the toes present the disturbances of acroparesthesia.

The symptoms of this vasomotor neurosis are intermittent, there being intervals entirely or nearly free from manifestations. During the attacks the suffering may be very severe, especially late at night or in the very early morning hours. The sensations are described as numbness, stiffness, awkwardness or clumsiness of the fingers, which feel swollen. Objectively, the affected digits are often reddened and covered with perspiration, although their usual appearance is pale and cold. Hyperesthesia is occasionally present, but hypesthesia is more common. These patients give no evidence of involvement of the central nervous system, the vasomotor trophic neurosis being of a purely local character.

A transition to Raynaud's disease is represented by cases in which the affected fingers are discolored and cyanotic, with marked vasomotor disturbances and abnormal sensations of pain, touch and temperature. Other cases are related to scleroderma and to other vasomotor trophic neuroses, the acroparesthesia being followed by cutaneous atrophy and tightening of the skin over the finger-tips.

Treatment.—The condition is not dangerous, but obstinate and intractable, usually persisting for months and years. **Occupations** responsible for the disease—notably washing—**should be stopped** when possible, and the general health be brought up to the standard. Locally,

* See, Roussy, P., Boisseau, J., Oelsnitz, M. Les acro-contractures et les acroparalyses, leur nature, leur traitement. *Rev. de méd.* 1917, iv, 515.

massage, hot water douches, medicated or otherwise, electricity and similar measures, have been recommended and are often efficacious.

Pathology.—Suggestive of the possible pathology of this condition is an observation made at the Vanderbilt Clinic in which the author observed a herpes zoster of the forearm in a case of acroparesthesia. The herpes appeared in the more severely affected extremity. One can therefore consider the lesion as perhaps occurring in one of the posterior root ganglia.

INTERMITTENT CLAUDICATION

(*Dysbasia Angiosclerotica*)

Definition.—Intermittent claudication (intermittent limping), a term first applied by Charcot, denotes a syndrome due fundamentally to a pathological condition of the blood-vessels, usually of the arteries. The obtrusive clinical symptom is a transitory interference with active movement of the affected part, accompanied during these periods by pain and spasm. Muscular weakness or absolute disability with numbness and dysesthesiæ of various forms are the usual accompaniments of the paroxysm. Earlier descriptions alluded almost entirely to the involvement of the lower extremities, but we now know that almost any part of the body may be affected. The majority of patients are past forty.

Etiology.—The most common etiological cause of intermittent claudication in the reported cases seems to be abuse of tobacco, always in association with other injurious factors, such as abuse of tea, alcohol, exposure to cold, syphilis, flatfoot. Faulty nervous heredity was demonstrable in only one of the cases. The author has observed intermittent claudication very frequently among Russian Hebrews.

Intermittent claudication is occasionally observed in youthful patients free from all evidence of arteriosclerosis, but suffering, like the arteriosclerotic group, from functional changes of the nervous system, nicotin intoxication, etc. As a sequela of external injuries, such as contusions or fractures, inflammatory lesions of the soft parts, etc., the affected leg becomes cold and stiff, pulsation in the pedal arteries diminishes or disappears, and the characteristic attacks supervene when the patient attempts to walk, or sometimes, to a lesser degree, even with the limb at rest. The rare cases of this kind must be interpreted as resting upon a purely neurotic basis.

A predisposition to vascular spasms is undoubtedly created by a neuropathic diathesis, which must be emphasized as an important etiological factor in intermittent claudication. Nervous individuals usually have a very unstable vasomotor system, and minor stimuli suffice to induce vascular contractions of the peripheral vessels in particular. The pathological changes found have all confirmed the opinion that some form of arteriosclerosis is the basis of the symptom-complex. Less

marked changes have been observed in the veins, suggesting a phlebosclerosis. These changes are probably secondary to the arterial degeneration.

Symptomatology.—This condition is characterized by periodical attacks of limping, with rapid fatigue, and sensations of pain and stiffness in one or both legs after prolonged exercise. These manifestations are promptly relieved by rest. Arteriosclerotic changes, on a vasomotor basis, are usually considered as responsible for this condition, and this etiology is suggested by the designation of intermittent claudication as *dysbasia intermittens angiosclerotica* (Erb).

The existence of general arteriosclerosis alone does not suffice for the production of intermittent claudication. Among 800 arteriosclerotic individuals seen by Favre in the course of three years, only 8 patients presented the picture of intermittent claudication. In all of these 800 cases, *apokamnosis*, i.e., a pathological fatigability, and Goldflam's symptom were demonstrable. The last-named phenomenon consists in a remarkable pallor of the foot, sometimes also of the lower portion of the leg, observed when the patient is instructed, while in the horizontal position, to raise the extended leg repeatedly. Differences in the pulsation of the pedal arteries, or loss of pulsation, are frequently noted, and general arteriosclerosis is usually demonstrable.

Treatment.—In the treatment of *dysbasia angiosclerotica*, Pick recommends the combination of iodine and what may be termed **hypopressure respiration**, as follows: Fluid iodine is transformed into a dry vapor which the patient is made to inhale through a mask which hermetically closes off the face and thereby renders inspiration more difficult. It is claimed that this method insures a more active blood supply and thereby an improved nutrition of the extremity, while providing a more rapid drainage of the carbonic acid and of other waste products in the peripheral veins. The clinical picture is said to improve and the vasoconstrictor symptoms gradually to disappear as a result of the energetic oxidation due to this mode of treatment.

PERSISTENT HEREDITARY EDEMA OF THE LEGS

(*Milroy's Disease*)

Definition.—Persistent hereditary or congenital edema of the legs, first described by an American physician, W. F. Milroy, in 1855, is characterized, as its name implies, by an edema of the lower extremities. It is not due to obstruction in the veins or the lymph-channels, and is not of constitutional origin. The edema is chronic.

Etiology.—Milroy's disease is a rare familial trophoneurosis, also known as persistent hereditary edema of the lower limbs. Either parent, although free from the disease, may transmit it to the offspring of both sexes. A neuropathic family history can usually be elicited, showing the occurrence of epilepsy, inebriety, feeble-mindedness or other

signs of constitutional inferiority or degeneration among the patient's relatives. These conditions may likewise be found in the patient himself.

Three possible local causes have been suggested: (1) venous obstruction or thrombosis; (2) lymphatic obstruction; (3) errors in the behavior of the blood-vessels or lymphatics without there being any actual obstruction to them, that is, a *vasomotor neurosis*. So far no changes have been demonstrated in the lymphatics or veins. In the author's opinion the condition may be regarded as a central vasomotor or trophic disturbance.

Symptomatology.—Edema, without a local or general cause to account for it, in a subject giving evidence of faulty heredity, should arouse the suspicion of Milroy's disease. These cases are characterized by a sudden transition from the edematous to normal tissue at the level of Poupart's ligament or at the knee-joint in the more restricted cases. The nature of the process is a matter of controversy. There is no venous or lymphatic engorgement. True objective sensory disturbances are absent, nor are there changes of the muscles to electrical stimulation; there are no atrophies. The sluggish reaction of the muscles to electrical stimulation, occasionally observed, is due to the resistance offered by the thickened and abnormally dry skin.

The characteristic findings are well described in the following succinct summary of Hope and French:

- (1) Restriction of the edema entirely to the legs.
- (2) Absence of any traceable cause for the edema, general or local.
- (3) Strong family predisposition to the complaint.
- (4) Painlessness of the pale swollen leg (apart from the acute attacks).
- (5) Absence of constitutional symptoms.
- (6) Sharpness of limitation of the upper level of the edema.
- (7) Incidence in both males and females.
- (8) Permanence of the edema once it is established.

Milroy's disease has been compared by some observers to the muscular dystrophies. The latter may develop in infancy or about puberty, or they may appear in adolescence, or, more rarely, in middle life. Both of these syndromes are distinctly familial. Similarly, it seems that tropho-edema of the legs may be congenital and present at birth, as in Milroy's cases. Or it may be hereditary and only develop later at a regular period after birth (puberty), as in Meige's cases, or at a variable period after birth. The form of hereditary transmission is comparable to that of Huntington's chorea. The subject of one of the author's observations was a young woman in whom the condition became markedly apparent at the age of thirty years. There was a family tendency to edema of the lower extremities, together with a neuropathic family history.

Treatment.—Treatment seems to have no effect on the condition.

For successful symptomatic treatment, constant and properly applied **bandaging of the extremities** is advisable. The methodical and early application of bandages is necessary for limiting the edema. Pa-

tients can thus be made comfortable. If allowed to extend, the swelling may reach proportions which seriously restrict activity.

Prognosis.—The disease has no influence upon longevity.

TROPHEDEMA

Definition.—Trophedema is a chronic neuropathic edema occurring in segmentary distribution associated with a painless hardening and pallor of the skin, not due to cardiac or renal disease. The condition of edema of trophic origin can be considered only when complicating conditions are excluded.

Etiology.—Trophedema, a disease, or rather a syndrome, first described by Meige in 1898, is a condition of somewhat obscure origin, the anatomicopathological basis of which still remains to be elucidated. What little is known concerning its etiology strongly suggests a part played by the nervous system in the form of a disturbance of the sympathetic system or possibly trophic centers within the spinal cord. A relation seems to exist between chronic trophedema and certain vasomotor disturbances, mental affections, hysteria and epilepsy. A coincident trophedema has been noted in the paralyzed parts, in a few cases of hemiplegia and paraplegia. As a rule, however, all evidences of an organic disease of the nervous system are absent. On the basis of a few isolated observations on trophedema of apparently traumatic origin it has been interpreted as developing through the intermediation of an ascending sensory neuritis acting upon the cells of the corresponding spinal ganglion and, from here, on the constituents of the neighboring sympathetic ganglia.

Some instances of chronic trophedema have been observed in which the swelling was associated with disturbances of the ductless glands in the form of more or less marked myxedema or acromegaly. Infection, although usually merely in the form of a preliminary attack of one of the exanthemata, has occasionally been held responsible, and in a few cases the onset of the swelling is said to have been preceded by diffuse pain and a rise of temperature.

The disease may be acquired, supervening at a variable age, usually about puberty, or it may be hereditary and congenital, the infirmity being present at birth. In either case, the trophedema is to be interpreted as a familial disease. It has been noted that in the hereditary cases the disease tends to be transmitted through the maternal line, and that in a general way women are especially susceptible to chronic trophedema. These cases must be carefully differentiated from *adiposis dolorosa*.

Symptomatology.—As a rule, trophedema first manifests itself by the immediate appearance of the swelling, without inflammatory prodromata, although the establishment of the edema is sometimes preceded or accompanied by severe neuralgic pains or spasms. The evolution of the edema may extend over an extremely variable period of time—from a

few days to a year. The swelling develops progressively, as a rule, advancing at a more or less gradual rate; in other cases, acute successive attacks occur in the affected region, each leaving an increasingly pronounced and persistent swelling. In the absence of a demonstrable cause, extensive regional edemas develop and cause a fibrous hardness,



FIG. 12.—A FORM OF PSEUDO-ELEPHANTIASIS.

Marked changes of trophic nature (secondary), due to mechanical obstruction.

covering the entire limb. In other cases, the swelling is, from the start, very hard and non-depressible.

The disease has its site of predilection in the lower limbs, extending to a variable level, up to the knee or groin. One side alone or both may be attacked symmetrically. The genitals and the lower portion of the abdominal wall escape, as well as, usually, the toes, although the instep of the foot is apt to be involved. Trophedema may likewise affect the upper limb, especially the fingers, the hand and the forearm. The face and the trunk are only exceptionally attacked. A segmentary distribution of the edema is emphasized by many observers.

The appearance of chronic trophedema resembles that of elephantiasis at first sight, and the disease is therefore sometimes designated as neuro-arthritic pseudo-elephantiasis. The lower limb, especially, may assume startling proportions and present enormous deformity. The skin in recent cases remains smooth, of a normal color, without venous or lymphatic varicosities, but it is adherent to the subjacent tissues and cannot be folded or indented. Later on the skin becomes thickened, while retaining its normal color and general sensibility. The disease is slowly progressive or remains practically stationary, and in all cases it is prolonged over several years without disturbance other than the impaired motility caused by the disproportionate enlargement of the affected leg.

Diagnosis.—As stated above, trophedema is often confused with *elephantiasis*, due to a similarity in the symptoms and manifestations of the two conditions.

Elephantiasis is a disease of tropical and subtropical countries, marked by chronic hypertrophy of the skin and subcutaneous tissue, leading to enormous enlargement of a particular part of the body, generally one, and in rare cases both of the lower limbs. The face is sometimes the site of the disease.

The condition is a pachydermia, with thickening and deformity of parts of the body, the skin as well as the underlying tissues being affected, due to local circulatory disturbances, such as chronic inflammation of the lymphatics and veins as well as of the cellular tissue. The ultimate cause of the changes is the plugging of the lymph-channels of the affected part, which is usually due in tropical countries to the *Filaria sanguinis hominis*, also known as the *Filaria bancrofti*. There is also a bacterial form of elephantiasis, due to streptococci. Lymphatic obstruction in other cases is the result of severe or recurrent inflammation, as in erysipelas, phlegmasia dolens, protracted eczema or other factors interfering with the lymphatic circulation.

The accompanying photograph illustrates a form of elephantiasis due to mechanical obstruction within the pelvis, disturbing the local venous lymphatic circulation to a marked degree. It is likely that the vasomotor and trophic centers are secondarily involved in cases of this kind and that they may be in part responsible for the intense nutritional disturbances. (Fig. 12.)

Treatment.—Therapeutic measures are not promising, but an attempt may be made with thyroid medication. Massage and elastic compression, with rest in bed, sometimes temporarily relieve the swelling.

NEUROFIBROMATOSIS

(von Recklinghausen's Disease)

Definition.—Neurofibromatosis, or von Recklinghausen's disease, is condition characterized by the occurrence of tumors in the form of

multiple neuromata, or circumscribed hyperplasia of the fibrous structure of the nerves. These growths may be in the form of soft nodules, sometimes pedunculated, of varying size, location and number. When the growths are very large and generalized, we speak of the condition as elephantiasis neuromatosa. The associated brown pigmentation of the skin, local or general, is characteristic of the condition.

Etiology.—On the basis of observations in cases of Recklinghausen's disease, some writers believe that the *endocrine glands* are often involved in this condition. The disturbances of the internal secretions may be due to involvement of the *hypophysis* or the *sympathetic system*, the affection of these representing a part of the congenital systemic disturbance, to which condition Recklinghausen's disease is attributed by some observers. A patient, a boy of nine years, observed by Lier, presented a well-marked dystrophia adiposogenitalis, in addition to the fibromatosis. The x-ray and ophthalmoscopic findings suggested a tumor of the hypophysis.

The disease may have a congenital basis dating to embryonic life.

Upon the basis of their observation of two cases of family neurofibromatosis (in mother and son)—one of which was associated with acromegaloid deformities—two recent French observers, Roubinovitch and Regnault de la Sourdière, interpret these and similar cases as due to a congenital dystrophy of the nervous and cutaneous ectodermic tissues, with a variable localization in different individuals, but of identical nature. Although existing at the time of birth, the condition is sometimes not manifested until a more or less advanced age.

Symptomatology.—Neurofibromatosis, or Recklinghausen's disease, is characterized by cutaneous or subcutaneous fibrous tumors, pigmentation of the integument, and stigmata of degeneration, or perhaps more properly speaking, anomalies of development, both physical and psychological. In those cases where the tumors develop on a peripheral nerve or on a spinal root, they give rise to symptoms corresponding to their localization. They have been known to occur in the spinal canal and to cause paraplegia through compression of the cord. Fibrous growths of this description, developing at the origin of cranial nerves—especially the acoustic nerves—have a sarcomatous structure and are associated with symptoms of brain tumor. The facial and trigeminal nerves are less frequently the sites of the growths.

Preiser and Davenport, in a careful study of the literature of 243 cases, found that feeble-mindedness occurred in 7.8 per cent. The disease may recur without a break for generations. A strong hereditary tendency is present. The above-named observers outlined charts showing that in families disposed to the disease, where there were two or more children, 43.5 per cent. of the offspring were affected with some form of neurofibromatosis. Furthermore, there is a tendency for the growths to affect the same parts of the body in certain families, and tumors of unusual localization appear in corresponding parts of the body in the case of members of the same family, as, for instance, in the case of brothers.

Clinical Varieties.—Aside from typical neurofibromatosis, there are also *incomplete* or *abortive* forms, in which only one of the cardinal symptoms is present, namely, either pigmentation or fibroma, which may be not infrequently combined with mental defects or skeletal anomalies.

Berg describes numerous clinical cases of epilepsy and idiocy, associated with various cutaneous changes, in the form of facial warts, showing a close relationship in part with neurofibromatosis, and in part with tuberous sclerosis. The cutaneous phenomena and the cerebral disturbances are interpreted as parallel manifestations, and, on the basis of these cutaneous changes, it is possible to differentiate, during life, a certain group of pseudo-epileptic individuals from cases of genuine or idiopathic epilepsy.

Treatment.—The treatment consists in obtaining relief by the **surgical removal of the growths** when these are so situated as to be within operable areas.

Bearing in mind the possible endocrine dysfunction as a factor, **glandular therapy**, especially the administration of **suprarenal gland**, may be employed.

Recently injections of Mendel's **fibrolysin**, 2 c.c. every few days, have been favorably reported.

TROPHIC DISTURBANCES FOLLOWING ACUTE LESIONS OF SPINAL ORIGIN

Acute Decubitus.—**DEFINITION.**—Acute decubitus, or "acute bed-sore," is really a trophoneurosis, probably of central origin, manifested as a slough or breaking down of areas of skin and of the underlying soft parts.

ETIOLOGY.—This trophoneurotic process was attributed by Charcot to an irritation of the posterior central portion of the gray matter of the cord, because it does not occur in infantile paralysis nor in acute or chronic spinal paralysis of adults. He also observed decubitus occurring as a sequela of lesions of the cauda equina, and he admitted a decubitus of peripheral origin. Déjerine and Leloir, as well as other observers, showed that neuritic lesions are invariably present in decubitus, supervening as a sequela of severe articular rheumatism, tabes, hemiplegia, and multiple sclerosis. As pointed out by Déjerine, the degeneration of the nerves and the subsequent decubitus are probably the result of interruption of the trophic influence in the central nervous system, external pressure simply playing the part of an occasional determining cause.

SYMPTOMATOLOGY.—Decubitus is known to occur in the apoplectic stage of cerebral hemorrhage and, when due to cerebral lesions, the slough is situated on the buttock of the opposite side, whereas in spinal lesions it makes its appearance on the middle line at the level of the sacrum. In unilateral myelopathies, it develops, as might be expected,

on the anesthetic side. Sloughs in many cases occupy the greater trochanter, the internal aspect of the knees, or the heel, namely, the points exposed to pressure, but they also occur on the abdominal walls and the dorsal surface of the foot. Irrespective of its cerebral or spinal



FIG. 13.—ACUTE DECUBITUS.

Showing deep ulcerative changes of the cutaneous and subcutaneous tissues of buttocks, thighs, and feet. Secondary to myelitis.

origin, decubitus is associated with very extensive parenchymatous neuritis of the nerves of the skin of the affected part. The accompanying photographs show unusually extensive trophic changes of the subcutaneous tissues in a case of transverse myelitis which came under ob-



FIG. 14.—ACUTE DECUBITUS. (Same case as Fig. 13.)

servation in the author's wards at Montefiore Hospital. (See Figs. 13 and 14.)

Arthropathies.—ETIOLOGY.—Lesions of the *spinal cord* (tabes, syringomyelia, etc.) and of the *brain* (for example, cerebral hemiplegia) may be attended by trophic affections. Tabes is well known to be accompanied by trophic degenerative changes, especially in the joints, with

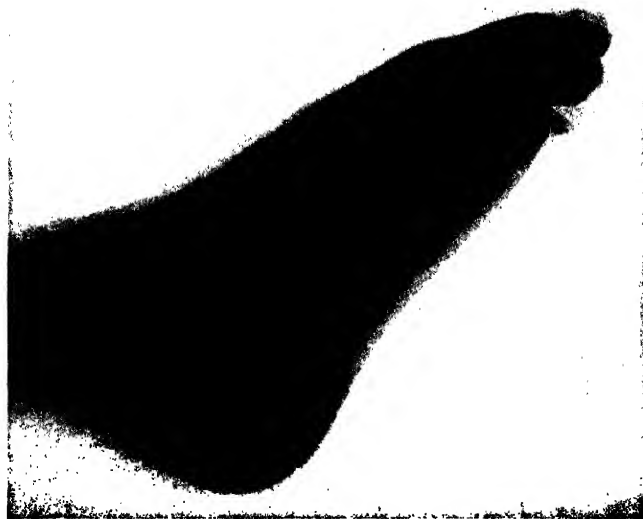


FIG. 15.—TABES DORSALIS, SHOWING OSSEOUS CHANGES IN THE FOOT.

Very advanced, irregular, diffuse bone atrophy, moderate bony arthritic changes. Marked infiltration of soft tissues with fibrous degeneration.



FIG. 16.—TABES DORSALIS, SHOWING CHARCOT JOINT IN THE FOOT.

Marked irregular diffuse bony atrophy associated with bony arthritic changes, especially along tarsus. Posterior portion of os calcis separated and displaced upward by the tendo achillis.

hypertrophy as well as erosion of the articular cartilages and sudden large serous effusions into the joint cavity, sometimes resulting in dislocation. These trophic changes are usually summed up together under the name of *Charcot's joint disease*. (Figs. 15, 16 and 17.) In the author's experience the vertebræ are not uncommonly the seat of these

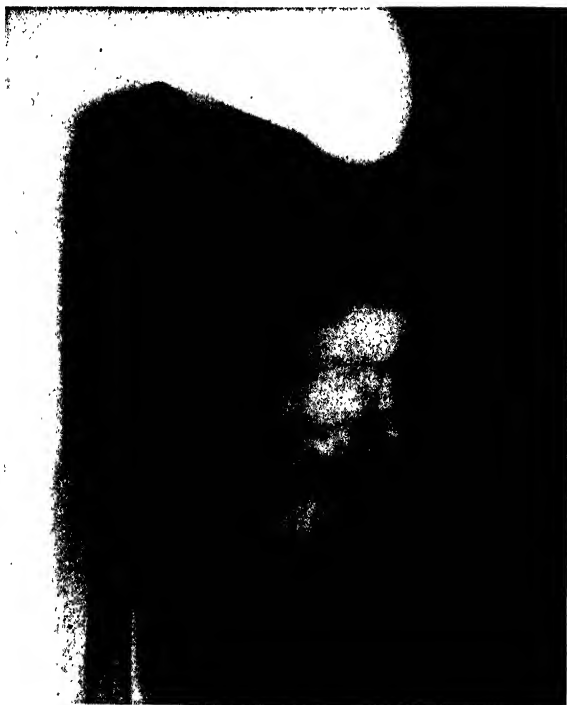


FIG. 17.—TABES DORSALIS, SHOWING OSSEOUS CHANGES IN THE SHOULDER.
Irregular bony atrophy. Slight arthritic changes in shoulder.

arthritic changes in tabes. (See Figs. 18, 19 and 20.) Perforating ulcer of the foot and abnormal brittleness of the bones are other trophic manifestations of this disease. Painless spontaneous fracture is common. (Fig. 21.) In syringomyelia, the loss or impairment of pain-sense becomes gradually associated with evidence of trophic disturbance through nutritional changes in the bones, muscles, and skin. The shoulder-joints, elbow-joints and wrist-joints become swollen, filled with fluid, and absorption of the articulating surfaces takes place, the condition in the

upper extremities in this disease being practically identical with that seen in the joints of the lower extremities in certain cases of locomotor ataxia. (Figs. 22, 23, 24 and 25.)

The development of the osteopathies and arthropathies in *tabes* and *syringomyelia*, as has been interpreted by Turney, is determined as follows:



FIG. 18.—CHARCOT SPINE.

Irregular destruction of body of 4th lumbar vertebra, associated with abundant new bone formation along both lateral aspects. The 4th lumbar vertebra prolapsed anteriorly (spondylolisthesis), thereby giving lumbar spine an abnormally short appearance.

(1) The fundamental change is the same in both, and consists of an atrophy of the bone.

(2) This is a true trophic lesion, to be distinguished from that due to disuse. It is produced reflexly by irritation and is strictly analogous to arthritic atrophy of the muscle.

(3) The general type which is assumed by an arthropathy is determined by local causes.

Aside from tabes and syringomyelia, certain other diseases of the spinal cord usually give rise to trophic disturbances. *Anterior poliomyelitis*, which is free from a tendency to bed-sores or to trophic changes in the skin, sometimes may lead to *acute atrophy of the bones*.

Arthropathies have been reported following *Pott's disease* of the spine, *acute myelitis* and, very exceptionally, after progressive muscular

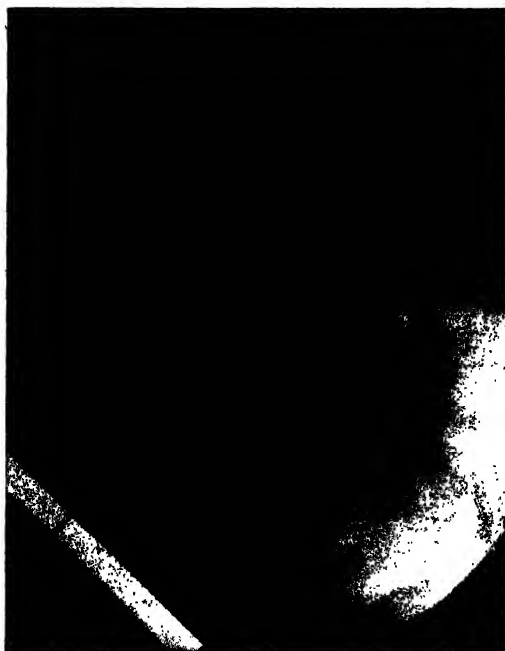


FIG. 19.—CHARCOT SPINE IN TABES DORSALIS.

Involving 2nd and 3rd lumbar vertebræ. Falling in of left side of both vertebræ, owing to irregular bone destruction in that region, associated with great amount of new bone formation. The latter resembles large bony bridge between vertebræ, resulting in marked midlumbar curvature. All the vertebræ show more or less marked deformity. (All of the roentgenograms were made by Doctor Thomas Scholz.)

atrophy and amyotrophic lateral sclerosis. The author's experience is in accord with that of Turney, who calls attention to the fact that although trophic defects are not frequent in multiple sclerosis, some degree of vasomotor disturbance is more common than is generally supposed.

Fundamentally the changes observed in acromegaly are of trophic origin; here the unquestionable rôle of the endocrine glands bespeaks

a probable participation of these glands in other forms of osseous change. (See Figs. 26 and 27.)

The hypertrophic and atrophic arthritic changes seen in *arthritis deformans* are chronic joint processes of unknown etiology. The changes in the joints resemble those found in Charcot's disease, but the condition is not associated with tabes or with any other spinal affection. The trophic disturbances affect the articular cartilages, the ends of the bones, and the synovial membranes. There is a difference of opinion as to

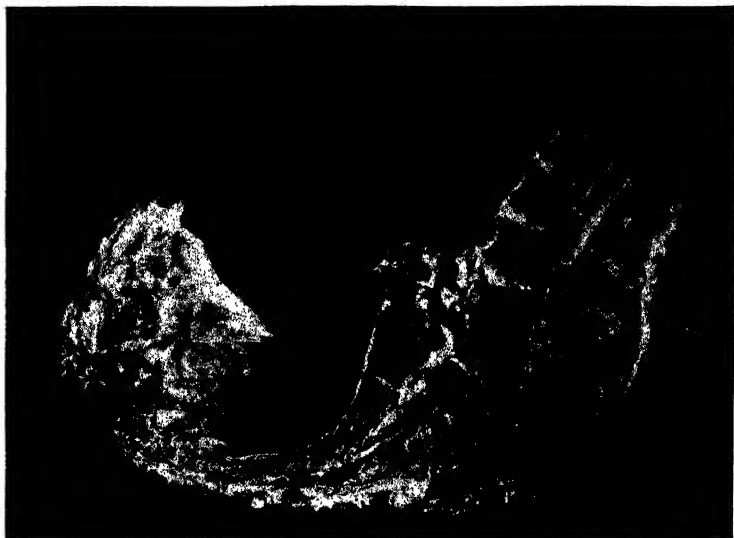


FIG. 20.—SAGITTAL SECTION THROUGH LUMBOSACRAL VERTEBRAL COLUMN, SHOWING CHARCOT TYPE OF DEFORMITY IN A TABETIC.

the infectious, bacterial character of this disease or its causation by primary nerve lesions. Observation of extensive material at the Montefiore Hospital, in New York, leads the writer more and more to the point of view that these arthritic changes have their origin in the central nervous system. (Fig. 28.)

VASOMOTOR AND TROPHIC DISTURBANCES IN TRAUMA OF PERIPHERAL NERVES

Etiology.—Our knowledge of vasomotor, secretory and trophic disturbances has been very much extended by the war studies of injuries to the peripheral nerves. Much error will be avoided in future by the appreciation of the fact that the disturbances are fundamentally due

to *injury of the nerves*, and not to the accompanying vascular injury. There is abundant evidence to show that nerve injury *per se* may be responsible for profound vasomotor disturbances. As shown in many cases of gunshot wounds in which there is only partial division of the peripheral nerve, we find vasomotor and trophic symptoms as obtrusive manifestations, apparently as a result of nerve irritation.



FIG. 21.—TABES DORSALIS, SHOWING PATHOLOGICAL FRACTURE THROUGH LOWER TIBIA AND FIBULA.

Marked splintering of fragments. Absence of any callus though fracture of longer standing. Very advanced irregular bony atrophy.

Experiences in the World War with vasomotor disturbances appearing in paralyses and contractures of apparently reflex character, lead a number of French observers (Babinski, Froment, Heitz) to regard these manifestations as independent of vascular or peripheral nerve lesions. Their distribution and variability suggest reflex stimulation of the sympathetic ganglionic centers, perhaps also of the bulbospinal centers.



FIG. 22.—SYRINGOMYELIA, SHOWING TROPHIC OSSEOUS CHANGES IN LEFT SHOULDER.

Also enophthalmus and narrowing of left palpebral fissure, and spinal miosis of left eye due to involvement of the sympathetic ciliospinal center at 8th cervical and 1st dorsal segment of the spinal cord.



FIG. 23.—SYRINGOMYELIA, SAME CASE AS IN FIG. 22, SHOWING TROPHIC OSSEOUS CHANGES IN LEFT SHOULDER AND VERTEBRÆ.

The great variability and difficult interpretation of the vasomotor, secretory, and trophic disturbances associated with traumatism of the peripheral nerves are emphasized by Stopford, in his recent report to the British Medical Research Committee. Upon the basis of a careful examination of an amputated leg, in the case of a young man of twenty-three years who had fairly recently (about eighteen months prior to the amputation) received a severe injury to the nerve supply through a gunshot wound of the thigh, he very correctly concludes that the vascular



FIG. 24.—ROENTGENOGRAM OF SAME CASE OF SYRINGOMYELIA, SHOWING MARKED UNDERDEVELOPMENT OF RIBS AND OF BODIES OF VERTEBRÆ.

Also trophic changes in bodies of vertebræ (at points of arrows).

changes in the form of endarteritis localized in the distal part of the limb, increasing in severity toward the periphery, are secondary to the peripheral nerve lesion. The vascular changes caused by the nerve injury lead to severe ischemia, which in its turn is responsible for nutritional changes in the muscles, bones, joints, and skin. In other words, the irritative effect of peripheral nerve injuries upon the blood-vessels results in changes in the walls of the arteries supplied by the damaged nerves, in the form of a more or less marked narrowing of the vascular lumen. The author is in accord with Stopford in the con-

viction that there is enough clinical and pathological evidence to indicate that vascular changes produced by an irritative nerve lesion are the real cause of the so-called "trophic disturbances," and that the prompt relief of this irritation becomes an imperative measure for the prevention of complications, such as thermalgia, acute decubitus, and other nervous sequelæ of peripheral nerve injuries.

Tinel emphasizes that all trophic and vasomotor disturbances are more frequent and severe in neuritic irritations than in simple division

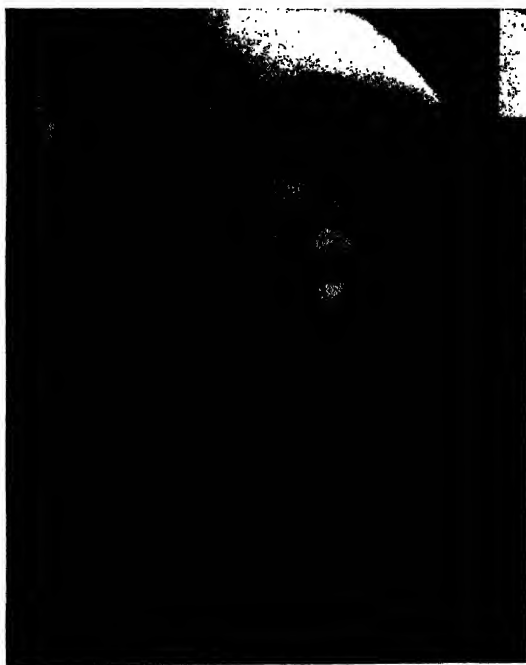


FIG. 25.—SAME CASE OF SYRINGOMYELIA, SHOWING CHANGES IN BODIES OF VERTEBRÆ AND SCOLIOSIS.

of a nerve. The only cause capable of producing equally severe trophic disturbances, as neuritic irritation, is arterial obliteration, giving rise to ischemic paralysis. Vascular lesions should, therefore, always be looked for in the presence of considerable trophic disturbance, especially as they often co-exist with nerve lesions and modify or exaggerate the clinical picture.

Symptomatology.—*Trophic disturbances* are usually absent or very slight in cases of simple nerve compression or interruption of nervous conductivity, whereas they are practically constant in the presence of nerve irritation and inflammation. A variety of disturbances are noted

in the integument, in the form of glossy skin, which is the most common, abnormality, in the secretion of the skin, either abnormal moisture or a peculiar dryness of the skin being noted, sometimes associated with cutaneous desquamation in large shreds, or again in the thickening and hardening of the integument.

Vasomotor disturbances are practically constant in all nerve lesions, peripheral or central. In relatively rare cases there is *pallor of the integument*, coinciding with dryness and thickening of the skin, espe-



FIG. 26.—ACROMEGALY, SHOWING MARKED Burring OF THE TIPS OF THE TERMINAL PHALANGES.

Also broadening of metacarpal bones and phalanges. Some osseous arthritic changes at bases of phalanges.

cially on the palms of the hands and the soles of the feet. *Cyanosis* and *redness* of the affected parts are much more common, the former being most frequently observed as a manifestation of vasomotor paralysis affecting the vasoconstrictor apparatus in the paralyzed area. *Redness of the skin*, on the other hand, is met with especially in cases of mild neuritic or neuralgic irritation without paralysis. It is most marked in causalgia and is usually associated with *excessive perspiration*. In all probability it constitutes a phenomenon of active vasodilatation.

Edema is sometimes observed in cases of nerve injury as a result of posture and inactivity, but in other cases it follows upon a neuritic irritation and may be present to a considerable degree. Both cyanosis

and edema are very often due to vascular lesions combined with the nerve injuries.

True ulcerations are very rare in the course of peripheral nerve lesions and almost without exception require a special determining cause, the nerve lesion acting as a predisposing factor through the nutritional disturbances induced by it.

Thermic disturbances are noted in the paralyzed limbs, in the form of diminution or elevation of the local temperature. A permanent rise

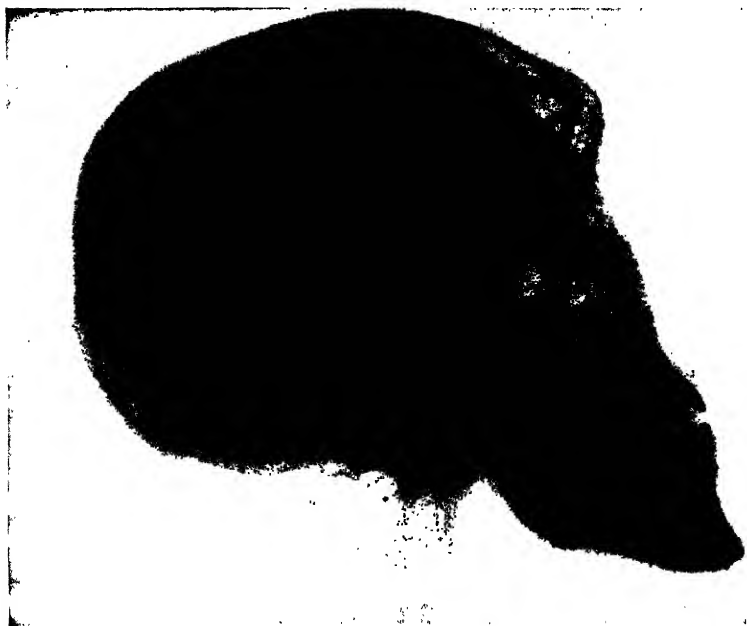


FIG. 27.—SKULL SHOWING OSSEOUS ELONGATION AND MARKED AËRATION OF SINUSES.

of temperature, with permanent vasodilatation and redness of the skin is noted only in certain mild neuritic irritations. Lowering of the temperature, which is very common, is more apparent than real, being due to a diminished circulation on exposure to the air. Considerable and permanent chilling of a limb is usually the result of vascular lesions associated with the nerve injury, and is combined in these cases with chronic cyanosis, edema, and progressive fibrous infiltration—in other words, with the features of progressive ischemia.

Trophic disturbances through nerve irritation may also affect the deeper layers, namely, the aponeuroses, tendons, synovial sheaths, bones and joints.

Concerning the trophic action of nerve fibers after trauma. Meige and

Athanasio-Benisty are inclined to think that an irritation of the nerve-trunk in the wound is transmitted by the centrifugal fibers to the sensory corpuscles of the skin, which through being continually irritated in this manner, give rise to severe pain, either spontaneously or as the result of external causes, such as temperature variations or circulatory changes. These causes affect the vascular tension in general and more



FIG. 28.—CASE OF ARTHRITIS SUGGESTING SECONDARY SYMMETRICAL INVOLVEMENT OF TROPHIC CENTERS.

particularly the numerous small capillaries in the papillæ of the skin, the glomeruli of the glands, and the sensory corpuscles. All changes of vascular tension react on certain fibers of the injured nerve which are neither motor nor sensory but take a centrifugal course and are distributed to the various layers of the skin and subcutaneous tissues. These centrifugal fibers pass either to the periphery of the nerve trunk or along its sensory fibers, probably also, to a great extent, to its most important nutrient arteries. Some of these fibers have probably a *trophic* action. As in the case of vasomotor fibers, their irritation causes

active vasodilatation in a succession of waves, considerable rise in local temperature, and atrophy and impairment of nutrition of all the tissues.

A *general atrophy* may be found, as shown by Babinski and Froment, especially when the wound is situated on the hand. This can be clearly seen in the tapering fingers. A comparison of the skiagrams of the two hands in Figs. 5 and 6 shows *decalcification of the skeleton*, which has been observed by a large number of writers, after traumatism of the skin. It is best seen in the extremities. In the *x-ray* plate, the skeleton of the affected hand appears to be more distinct and the outline of the bony trabeculae clearer and more delicate, while the epiphyses and joint surfaces are less distinct than on the sound side. The same appearances are to be found, but to a lesser degree, in the bones of the forearm.

There is almost always a certain degree of *fibrotendinous* or *muscular contracture*. The position of the contractures varies; they may be seen in the joints of the fingers, wrists, elbow, hips, foot and ankle, but they are most marked in the finger joints, which are frequently enlarged and painful like those of chronic rheumatoid arthritis. They develop rapidly and explain, to a certain extent, the limitation and difficulty in the movements of the joints. Although they are hardly ever absent in reflex contractures and pareses, their intensity varies. On the other hand, they are exceptional in cases of hysterical paralysis or contracture.

Nonne's observations on war injuries of peripheral nerves, comprising 152 cases, are worthy of study. In the microscopical examination of resected nerve segments, portions of projectiles were frequently encountered in cross-sections, and bone-splinters were not uncommon. In cases of clinically severe or moderately severe paralysis, the nerve is sometimes found intact at the time of operation, so that a concussion of the nerve-trunks must be assumed, associated with degenerative processes in the nerve. Very commonly, not all the motor functions governed by the injured nerve-trunk are lost or impaired. The same frequency of partial involvement can be established for the sensibility. Painful neuralgias in these cases are rarely observed, so that the sensory fibers seem to be considerably more resistant to the sequelae of traumatism than are the fibers transmitting motor and electric stimuli. What is true for the motor and sensory conduction is likewise applicable to the behavior of the electric irritability, which means that the change in electric reaction frequently affects only a part of the paralyzed nerve.

Trophic disturbances were found to be by no means rare in the war injuries of peripheral nerves, in the form of vasomotor disturbances in the paralyzed parts: cyanosis, cold, pallor, hyperhidrosis and anhidrosis, trophic changes of the nails, trophic disturbances of the epidermis, occasionally the formation of vesicles. Ulcerations were not observed, at least not in cases uncomplicated by frost-bite. Cases of a brown discoloration and hypertrichosis of the entire locally injured extremity have been reported by Oppenheim, who also speaks of general vasomotor and secretory disturbances after local war traumatism of

peripheral nerves, explaining these disturbances by a general traumatism of the central nervous system. These cases usually present general hyperhidrosis and reddening of the skin. This cannot be a frequent occurrence. Nonne was never enabled to observe it among his 152 cases, and is inclined to refer this phenomenon to psychic factors.

Babinski and Froment point out that *hypertrichosis* may be observed, not only in neuritis but also in reflex contractures and paresis. It is absent in purely functional contractures and paralyses. As a rule, the *nails* also show some changes in patients with reflex disorders, their growth being less rapid than on the sound side.

According to observations made in the course of the world war, by Villaret and others, hypertrichosis developing in the cutaneous territory of a wounded nerve generally coincides with hyperhidrosis and absence of the reaction of degeneration, and indicates an incomplete lesion of the nerve; whereas, on the contrary, hypotrichosis originating under the same conditions is usually accompanied by the reaction of degeneration and indicates a complete division of the nerve.

Trophic, vasomotor and secretory disturbances were observed by Steinberg in 52 of 70 cases, i.e., in more than 70 per cent. With special reference to *trophic disturbances*, these cases presented—aside from the ordinary muscular atrophies from disuse, or from destruction of the trophic center—bone atrophies demonstrable by the *x-ray*; a great variety of trophic disturbances of the skin, such as hyperkeratosis, which is relatively common in paralysis of the sciatic nerve; an abnormal vulnerability for higher degrees of temperature; an abnormally rapid growth of the nails, more frequently than the opposite, in the form of onycholysis, abnormal transverse and longitudinal furrowing, especially in affections of the radial nerve; furthermore, hypertrichosis, much more rarely hypotrichosis. Hypertrichosis is most frequently seen in ulnar lesions, most rarely in injuries of the sciatic nerve.

The *vasomotor disturbances* include edema, subjectively and objectively perceptible cold, and the presence of a purplish hue of the skin.

Secretory disturbances, such as hyperhidrosis, hypohidrosis, and anhidrosis have been observed, the first-named being the most common. These disturbances occur immediately or very soon after the injury in contradistinction to hypertrichosis, which is of late development.

The distribution, or severity, respectively, of all these trophic, vasomotor, and secretory disturbances is not demonstrably parallel with the severity of the motor or sensory paralysis.

Upon the basis of these observations, it is necessary to assume the existence of trophic nerves, whose mode of action must be considered as altogether different from that of motor or sensory fibers.

As a result of gunshot injuries in particular, the nerves may become imbedded in dense cicatricial tissue, and on operative exposure they are seen to be considerably altered, flattened or thickened, relaxed and flabby, the swollen and club-shaped ends consisting of neurofibromatous tissue. The resulting conditions are paralyses, sensory, vasomotor and trophic disturbances. Without operative intervention the majority of these

disturbances persist; spontaneous regeneration is to be anticipated only in the milder cases.

A case in point reported by Leszlenyi was one of trophic disturbance in the left radial region, due to chronic irritation exerted upon a cutaneous branch of the radial nerve by a foreign body, in the form of a projectile. The trophic and sensory disturbance in the territory supplied by the radial nerve and its cutaneous branches are explained as due to centripetal transmission, as well as by a sort of sympathetic disturbance of the nervous conductivity in the contralateral sphere of distribution of the nerve. The patient, a youth of eighteen years, had received a gunshot injury of the left upper arm; two bullets were removed, a third bullet was said to have entered, but could not be discovered. About two years later, a large gangrenous focus made its appearance on the left upper arm. There was also a sensory disturbance of the right hand, evidently of functional character, and secondarily superadded to the trophic disturbance of the left side.

Remote injuries of peripheral nerves through gunshot injuries were recently discussed by Strohmeier, who showed that nerves in tissues through which a projectile has passed may be so affected as a result of mechanical stretching as to result even in total loss of function. The injury gives rise to an endoneural hemorrhagic and lymphatic extravasate, from which, in its turn, a cicatrix results. In these cases, the nerve often still presents a macroscopically normal appearance, although in a number of instances it is club-shaped and swollen. An induration subsequently develops, with thickening and hardening of the nerve. Cicatrices of this kind must be treated by **exposure of the individual nerve-strands**, or by **excision**. Nerve lesions may also occur as a result of overdistention or rupture of the axis cylinders alone. In these cases, trophoneurotic disturbances may follow.

Lesions of the sympathetic system in war wounds of the peripheral nerves have been recently studied by Ferranni, based on 3 cases with vasomotor disturbances and edema of the hand, as the sequela of severe lesions of the radial and median nerves, and upon 2 cases of edema of the foot due to lesion of the sciatic nerve.

Edemas of the extremities may be observed as a result of traumatic lesions of the peripheral nerves (median, sciatic, internal popliteal) and sometimes of lesions of the spinal cord. When not dependent upon direct vascular lesions they are probably due to destructive or irritative lesions of the *sympathetic fibers* which accompany the fibers of the cerebrospinal system in the nerves. The fact that these edemas are most frequently observed in cases where the median nerve or the internal popliteal branch of the sciatic nerve are damaged is due to the fact that these nerves are more abundantly supplied with sympathetic fibers than are the others.

Sympathetic disturbances of the upper limb, due to lesion of the third and fourth dorsal roots, were observed by Tinel in the case of a soldier twenty-eight years of age. The clinical picture was characterized by a sensation of stiffness and tingling in the left hand, with

very pronounced vasoconstriction of all the small blood-vessels of the left upper limb, causing a waxy pallor of the hand, loss or extreme diminution of the radial pulse, and obliteration of the subcutaneous veins. This symptomatology in the present instance was evidently due to an irritative lesion of the third and fourth dorsal roots, which accordingly contain vasomotor sympathetic fibers passing to the upper limb.

Treatment.—The treatment of the vasomotor and trophic disturbances resulting from lesions of peripheral nerves is naturally fundamentally the same as that demanded for the restoration of the general function of the injured nerve. As Tinel says, every peripheral nerve suffering from a trauma shows a tendency toward regeneration, if the general health and condition of the patient are such that he can contribute toward this restoration. This remarkable inherent tendency of the nerves themselves to regenerate by growth of the axis cylinders into the periphery accounts for a very large number of spontaneous restorations in peripheral nerve injuries, and **surgical procedures** are directed toward bringing about this natural regeneration process which is effecting a coaptation of the two parts of a divided nerve. The object of the surgical procedure, when this form of therapy is undertaken, is to unite the proximal segment of the divided nerve which contains the axis cylinders to the distal portion of the nerve containing the nerve-sheaths; the proper surgical technic is of the utmost importance, and there are certain methods with which it is necessary for the surgeon to be familiar. Furthermore, an exact knowledge of the anatomy and physiology of peripheral nerves is essential. For example, the surgeon must know just where the nerve and its branches are given off, and what course they pursue; he must also necessarily know the nerve supply of the various muscles of the integument. Twisting and distortion of the nerve must be avoided; the fiber bundles within the nerve-sheath should be as far as possible approximated as before the section of the nerve occurred. Nerve-grafting has been frequently successful when extensive destruction of nerve-tissue has occurred. It appears, however, from a large statistical study of war wounds that less than 40 per cent. of peripheral nerve lesions require surgical treatment, whether by simple liberation of the nerve from scar tissue, or by suture of the segments of the severed nerve. While early surgical intervention, when this has been clearly indicated, is followed by the most satisfactory results in the form of early regeneration, experience teaches that surgical measures may be successfully undertaken many months after the injury, in cases where the regenerative process shows little or no tendency to spontaneous occurrence. Surgically, one of three measures may be undertaken to bring about restoration of peripheral nerve function after injury of the nerve. These are: (1) liberation of the nerve from the surrounding tissue which is crushing or pressing upon it; (2) suture of the severed nerve; (3) nerve-grafting. The latter procedure is resorted to in cases where the segments of the severed nerve are so separated as not to permit of direct suture of the parts. The material used for

grafting is usually a portion excised from some sensory nerve. The musculocutaneous from the lower extremity is often used.

Suture by division or splitting of the nerve is rather **less satisfactory**, although done in some cases. Various technical means are utilized when surgical procedures are employed to avoid the after-formation of cicatricial tissue. Among the more conservative methods of treatment are various forms of **electricity, mechanotherapy**—as: **massage and gymnastics, diathermia, radiotherapy, alcohol injections, ionization**, etc. All of these must be properly applied in selected cases.

VASOMOTOR AND TROPHIC NEUROSES OF NEURASTHENIC ORIGIN

Vasomotor disturbances are often noted in neurasthenic, or so-called vagotonic patients, individuals who are apt to complain of congestion of the head, or, still more frequently, of troublesome *blushing*. In the course of the examination, the face or one side of the face may become overspread with a diffuse redness, which sometimes extends in spots over the neck and chest. These patients are usually aware of the disturbance, and the mere idea of blushing is often a sufficient cause for its production. Erythrophobia, or fear of blushing, may become a very distressing and obstinate manifestation of vagotonia.

Related to vasomotor blushing are the phenomena of *urticaria factitia* and *dermographism*, conditions in which trifling irritation of the skin leads to well-marked and long-continued redness of the skin, sometimes with the formation of wheals. The influence of psychic factors and of intense emotion is well illustrated by the observation of a Russian writer, Agdshanianz, who recently (1913) described spasmodic ischemia as a cause of organic focal symptoms in a woman who presented vasomotor phenomena, with urticaria, pruritus and headache, as a sequel of severe emotional disturbances. A few days later, the patient was suddenly attacked by vertigo, diplopia, disturbances of deglutition and phonation, paresis of the left palatine velum, and general weakness. The urine contained 0.6 per cent. sugar. The patient's sister had died of polyneuritis with well-marked vagus symptoms, and she feared for her own life on account of the apparently similar illness. As soon as she had been positively reassured in this respect, the swelling promptly subsided, as well as the facial paralysis and diplopia; at the end of several days, the sugar likewise disappeared from the urine. The observer does not interpret the condition as due to hysteria, but as an angiospasm of functional character, which gave rise to a pure focal lesion with paralysis of a cranial nerve and glycosuria.

In a series of most instructive cases Lieut. Col. Hurst of the British forces demonstrated the trophic changes which may occur in purely functional paralysis, peripheral nerve injury. He showed local edema and peculiar trophic changes in the finger nails of the paralyzed hand or finger. Hurst attributes these changes to disuse only.

In a series of moving pictures shown at the meeting of the American Neurological Association (1919), Dr. Hurst demonstrated the fact of the immediate disappearance of these trophic changes, when, usually by suggestion or psychotherapy, the parts were restored to voluntary motion. However, in several cases of psychogenic origin under the writer's observation, the trophic disturbances, in the paralyzed limb, were, in the author's opinion, of more subtle origin. The author regards these changes as due rather to psychogenic factors. He does not consider simple stasis, due to local inactivity, sufficient to explain the often pronounced trophic disturbances observed in limbs paralyzed by purely psychic states.

CHAPTER XXIV

ANGIONEUROTIC EDEMA

By S. F. GILPIN, M.D.

Definition, p. 687—Etiology, p. 687—Symptomatology, p. 687—
Diagnosis, p. 688—Treatment, p. 688—Prognosis, p. 689—Pathol-
ogy, p. 689.

Definition.—This affection is characterized by circumscribed swellings of the subcutaneous or submucous tissues. It is known as giant urticaria, acute circumscribed edema, and Quinke's disease. It was first definitely described by Milton in 1876. Since then many cases have been observed in all parts of the earth. It may be hereditary, usually appears in those of a neuropathic constitution, and is often recurrent. It is commonly associated with gastro-intestinal disturbance.

Etiology.—The most prominent predisposing factors are heredity and exhaustion. Continued mental or physical exhaustion aids in the development of this vascular neurosis, yet the history of many cases gives no clew to the origin of this disease. The greatest number of cases appear in early adult life, from twenty to thirty-five years of age, though it has been seen in infancy. Both sexes are affected, though females more than males. A gouty tendency is present in some cases. Among the various factors thought to be exciting causes may be mentioned exposure to cold, emotional disturbances, puberty, the climacteric, and the action of toxic agents such as alcohol, tobacco and malaria. The malarial intoxication in some instances produced regularly recurring attacks. The attacks are most apt to occur during the latter half of the night when the activity of the circulation is the least. They are most apt to occur in the summer and winter and they may occur after slight traumatism.

Symptomatology.—The chief symptom of this disease is a characteristic swelling which comes on quickly and usually without warning. The swelling reaches its maximum extent in from one-half to two hours. It may shade into the surrounding tissue or it may be sharply defined. The color is usually whitish or waxy. The swelling is tense, not tender, does not pit on pressure, and rarely shows any purpuric discoloration. The center of the swollen area may extend one-half inch above the surrounding skin. The swelling lasts from a few hours to a day or two, and disappears as rapidly as it comes. The most usual locations are the face, lips, tongue, pharynx, genitals and extremities, though any part of the body may be attacked. The swelling may disappear from one part of the body to appear in another without any symmetrical or

anatomical relation. Several swellings may appear at the same time. The swelling may attain large proportions, thus giving rise to the term "giant urticaria," and nodular swellings as large as hen's eggs are sometimes seen. No objective sensory changes are found.

There are few subjective sensations in the swollen region, except a feeling of fullness, stiffness, and, in some cases, burning or itching. If the skin is scratched, urticarial wheals or stripes usually appear, or they may be associated with the attack or alternate with it. When the tongue, pharynx, larynx, stomach or intestines are affected, great local discomfort is caused, and, in the involvement of the pharynx, larynx, and tissues of the neck, as in a case recently under the writer's observation, death may ensue. Dyspnea, difficulty in deglutition, and gastro-intestinal colic are thus induced. Gastro-intestinal symptoms are very common and appear in one-half to one-third of the cases. The attack usually begins with a feeling of distress in the epigastric region, followed by nausea and gas distention. Cramps, vomiting and intense thirst ensue. The attack ends by a profuse diarrhea with watery stools. The secretion of urine may be increased during the attack, and it may contain albumin and hemoglobin. An effusion into the joints sometimes takes place. Cerebral symptoms as shown by lassitude, headache, somnolence, coma, or convulsions have been observed.

The attacks usually come on at irregular intervals of days and perhaps months. Between attacks the health is usually good. A blow is sufficient to induce and locate an attack in some cases and there seems to be a tendency for the attacks to recur constantly in the same locations. In some of the congenital cases the swelling may be permanent.

Diagnosis.—The diagnosis should present little difficulty. Some cases of chronic recurring articular rheumatism may bear a superficial resemblance to this affection, but it is always characterized by inflammation of the part and some general rise in temperature. The persistent blue and white edema of hysteria will be associated with the many symptoms, general bearing and susceptibility to suggestion so characteristic of hysteria. The edema of cardiac and renal disease is readily classified by a physical examination of the patient.

Treatment.—No specific treatment is known for this disease. The patient should be kept in the **best possible physical condition**, and an effort made to **remove all mental or physical causes** which may have a tendency to break down or lessen the patient's nervous resistance. With this in view, great care should be given to the regulation of the patient's **rest, diet, exercise and bathing**. Many drugs have been used, which fact alone shows that none of them are specific. **Atropin** is useful during the attack. **Strychnin** may be of benefit. Osler has seen good results from **nitroglycerin**. **Cold should be avoided**. With albumin or hemoglobin in the urine, rest and a **milk diet** should be insisted upon. **Calcium chlorid** or **calcium bromid** 9 to 15 grains (0.6 to 1 gram) three or four times daily has been recommended. Oppenheim speaks of two cases cured and one helped by the use of **quinin**. For the relief of the swellings **collodion** may be tried, or **compression**

by an elastic bandage. Edema of the glottis may require **scarification** or tracheotomy. **Spraying with a solution of adrenalin** may be of use. In a case recently under the writer's observation, in which the edema involved the pharynx, larynx, and all of the structures of the neck, intubation with the ordinary intubation tube was tried with no result; a flexible, firm, male catheter was then introduced through the larynx and into the trachea; artificial respiration was carried on, but with no effect, the patient dying, with no return of the respiratory function.

Prognosis.—There is little danger to life unless the neck, pharynx and larynx be involved, in which case the outlook is always alarming and at times fatal. There is a general tendency for the attacks to cease in advanced years, though they may last for life or may reappear after a long interval. If any inciting cause can be discovered in the individual case the outlook is improved.

Pathology.—The pathology of the disease is not well understood. Heidenhain believes that capillary cells play the chief rôle in lymph formation, and that morbid influences may lead to excessive secretion and production of swelling. Some think that chemical processes may have a causative relation. The familial forms of the disease are apt to appear early in life and show a wider range of symptoms.

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